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Braithwaite, W.R.;

Medical Technology Symposium, 1998. Proceedings. Pacific , 17-20 Aug. 1998
Pages:340 - 343

[\[Abstract\]](#) [\[PDF Full-Text \(20 KB\)\]](#) **IEEE CNF**

2 Medical information security: the evolving challenge

May, T.T.;

Security Technology, 1998. Proceedings., 32nd Annual 1998 International Carnahan Conference on , 12-14 Oct. 1998
Pages:85 - 92

[\[Abstract\]](#) [\[PDF Full-Text \(600 KB\)\]](#) **IEEE CNF**

3 Protecting personal data: can IT security management standards help

Iachello, G.;

Computer Security Applications Conference, 2003. Proceedings. 19th Annual , Dec. 2003
Pages:266 - 275

[\[Abstract\]](#) [\[PDF Full-Text \(273 KB\)\]](#) **IEEE CNF**

4 Development of a secure medical research environment

Alaoui, A.; Levine, B.; Cleary, K.; Mun, S.K.;

Information Technology Applications in Biomedicine, 2000. Proceedings. 2000 EMBS International Conference on , 9-10 Nov. 2000
Pages:44 - 49

10/17/341

[\[Abstract\]](#) [\[PDF Full-Text \(472 KB\)\]](#) [IEEE CNF](#)

5 Towards scalable authentication in health services

Gail-Joon Ahn; Dongwan Shin;

Enabling Technologies: Infrastructure for Collaborative Enterprises, 2002. WE 2002. Proceedings. Eleventh IEEE International Workshops on , 10-12 June 2002
Pages:83 - 88

[\[Abstract\]](#) [\[PDF Full-Text \(373 KB\)\]](#) [IEEE CNF](#)

6 E-knowledge in health care: a strategic imperative

Wickramasinghe, N.; Mills, G.L.;

System Sciences, 2002. HICSS. Proceedings of the 35th Annual Hawaii International Conference on , 7-10 Jan. 2002
Pages:1936 - 1945

[\[Abstract\]](#) [\[PDF Full-Text \(499 KB\)\]](#) [IEEE CNF](#)

7 Exploring the Moderating Effect of Trust and Privacy in the Adoption Application Service Providers in the Healthcare Industry

Randeree, E.; Kishore, R.; Rao, H.R.;

System Sciences, 2005. HICSS '05. Proceedings of the 38th Annual Hawaii International Conference on , 03-06 Jan. 2005
Pages:259a - 259a

[\[Abstract\]](#) [\[PDF Full-Text \(184 KB\)\]](#) [IEEE CNF](#)

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1 Anonymous E-prescriptions

Giuseppe Ateniese, Breno de Medeiros

 November 2002 **Proceedings of the 2002 ACM workshop on Privacy in the Electronic Society**

 Full text available: pdf(304.10 KB) Additional Information: [full citation](#), [abstract](#), [references](#)

This paper studies issues related to privacy protection of medical data, arguing that the topic is suitable for applied cryptographic research. We present the problem of medicine prescription privacy and describe a practical system that employs standard cryptographic techniques to achieve several improvements over current practices. We also introduce a very simple tool: Online group signatures which can be built via simple primitives implemented in commonly employed cryptographic libraries.

Keywords: medical information privacy, privacy-preserving cryptographic techniques, public-key cryptography

2 Privacy of medical records: IT implications of HIPAA

David Baumer, Julia Brande Earp, Fay Cobb Payton

 December 2000 **ACM SIGCAS Computers and Society**, Volume 30 Issue 4

 Full text available: pdf(819.71 KB) Additional Information: [full citation](#), [abstract](#)

Increasingly, medical records are being stored in computer databases that allow for efficiencies in providing treatment and in the processing of clinical and financial services. Computerization of medical records has also diminished patient privacy and, in particular, has increased the potential for misuse, especially in the form of nonconsensual secondary use of personally identifiable records. Organizations that store and use medical records have had to establish security measures, prompted pa ...

3 Can GRID services provide answers to the challenges of national health information sharing?

I. Bilykh, Y. Bychkov, D. Dahlem, J. H. Jahnke, G. McCallum, C. Obry, A. Onabajo, C. Kuziemsky

 October 2003 **Proceedings of the 2003 conference of the Centre for Advanced Studies on Collaborative research**

 Full text available: pdf(964.34 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#)

10/014.341

It has been widely recognized that one of the keys to cost reduction and service improvement in national health care lies in the integration of medical information system. Integration of information can not only improve care delivery today, but it can also help build research bases to enhance future care delivery. The question is how to achieve such integration? Imposing a single client software solution or common clinical terminology does not appear likely to happen. That lack of single softwar ...

4 Applications: A context-related authorization and access control method based on RBAC:

Marc Wilkens, Simone Feriti, Alberto Sanna, Marcelo Masera

June 2002 **Proceedings of the seventh ACM symposium on Access control models and technologies**

Full text available:  pdf(260.70 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#), [index terms](#)

This paper describes an application of authorization and access control based on the Role Based Access Control (RBAC) method and integrated in a comprehensive trust infrastructure of a health care application. The method is applied to a health care business process that involves multiple actors accessing data and resources needed for performing clinical and logistics tasks in the application. The notion of trust constituency is introduced as a concept for describing the context of authorisation. ...

Keywords: role based access control (RBAC), secure health care system, trust infrastructure

5 Full papers: Privacy-preserving data integration and sharing

Chris Clifton, Murat Kantarcioğlu, AnHai Doan, Gunther Schadow, Jaideep Vaidya, Ahmed Elmagarmid, Dan Suciu

June 2004 **Proceedings of the 9th ACM SIGMOD workshop on Research issues in data mining and knowledge discovery**

Full text available:  pdf(135.55 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#)

Integrating data from multiple sources has been a longstanding challenge in the database community. Techniques such as privacy-preserving data mining promises privacy, but assume data has integration has been accomplished. Data integration methods are seriously hampered by inability to share the data to be integrated. This paper lays out a privacy framework for data integration. Challenges for data integration in the context of this framework are discussed, in the context of existing accomplishment ...

6 Privacy, information technology, and health care

Thomas C. Rindfussch

August 1997 **Communications of the ACM**, Volume 40 Issue 8

Full text available:  pdf(561.04 KB) Additional Information: [full citation](#), [references](#), [citations](#), [index terms](#), [review](#)

7 Computer applications in health care (CAHC): Disclosure risk measures for the sampling disclosure control method

Traian Marius Truta, Farshad Fotouhi, Daniel Barth-Jones

March 2004 **Proceedings of the 2004 ACM symposium on Applied computing**

Full text available:  pdf(545.50 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#)

In this paper, we introduce three microdata disclosure risk measures (minimal, maximal and weighted) for sampling disclosure control method. The minimal disclosure risk measure represents the percentage of records that can be correctly identified by an intruder based

on prior knowledge of key attribute values. The maximal disclosure risk measure considers the risk associated with probabilistic record linkage for records that are not unique in the masked microdata. The weighted disclosure risk me ...

Keywords: data privacy, disclosure risk and sampling, microdata, statistical disclosure

8 Computer science in health and education: The use of smart devices in eHealth

John Fulcher
September 2003 **Proceedings of the 1st international symposium on Information and communication technologies**

Full text available: pdf(124.20 KB) Additional Information: [full citation](#), [abstract](#), [references](#)

Results from a field trial involving the use of USB iKeys as a secure access mechanism for remote access of patient medical records from a central server are reported. These are discussed within the context of eHealth generally, where technological considerations can easily be outweighed by concerns of patient privacy, security and confidentiality.

9 Information technology and dataveillance

Roger Clarke
May 1988 **Communications of the ACM**, Volume 31 Issue 5

Full text available: pdf(1.89 MB) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#), [index terms](#), [review](#)

Data surveillance is now supplanting conventional surveillance techniques. With this trend come new monitoring methods such as personal dataveillance and mass dataveillance that require more effective safeguards and a formal policy framework.

10 Security watch: The HIPAA-potamus in health care data security

Rebecca T. Mercuri
July 2004 **Communications of the ACM**, Volume 47 Issue 7

Full text available: pdf(64.80 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#) html(18.39 KB)

Regulations intended to improve health care data access have created new security risks along with headaches for patients and practitioners.

11 Information-rich commerce at a crossroads: business and technology adoption requirements

Robert G. Fichman, Mary J. Cronin
September 2003 **Communications of the ACM**, Volume 46 Issue 9

Full text available: pdf(108.80 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#), [index terms](#)

The day is approaching when most of our common transactions may be information-rich, but first an extensive supporting infrastructure must be developed in three areas: devices, networking, and trust.

12 Large systems: From privacy promises to privacy management: a new approach for enforcing privacy throughout an enterprise

Paul Ashley, Calvin Powers, Matthias Schunter
September 2002 **Proceedings of the 2002 workshop on New security paradigms**

Full text available: pdf(657.65 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#), [index terms](#)

Regulations and consumer backlash force many organizations to re-evaluate the way they

manage private data. As a first step, they publish privacy promises as text or P3P. These promises are not backed up by privacy technology that enforces the promises throughout the enterprise. Privacy tools cover fractions of the problem while leaving the main challenge unanswered. This article describes a new approach towards enterprise-wide enforcement of the privacy promises. Its core is a new framework for ...

Keywords: E-P3P, enterprise privacy management, privacy policy

13 Computer applications in health care (CAHC): Health Level-7 compliant clinical patient records system 

Jagbir S. Hooda, Erdogan Dogdu, Raj Sunderraman
March 2004 **Proceedings of the 2004 ACM symposium on Applied computing**

Full text available:  pdf(177.31 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#)

We present the design and implementation of a Health Level-7 (HL7)-compliant web-based clinical patient records system (CPRS). HL7 is one of the leading standards for exchange of clinical and administrative data among healthcare information systems. Since the passage of the Health Insurance Portability and Accountability Act of 1996 (HIPAA) by US government, the security of electronic medical clinical records systems is of paramount importance. HIPAA requires that various technical, physical and ...

Keywords: HL7, XML, clinical document architecture, medical informatics, schema, universal patient records, web-based application

14 Social Analyses of Computing: Theoretical Perspectives in Recent Empirical Research 

Rob Kling
January 1980 **ACM Computing Surveys (CSUR)**, Volume 12 Issue 1

Full text available:  pdf(3.98 MB) Additional Information: [full citation](#), [references](#), [citations](#), [index terms](#)

15 A web-enabled framework for smart card applications in health services 

Alvin T. S. Chan, Jiannong Cao, Henry Chan, Gilbert Young
September 2001 **Communications of the ACM**, Volume 44 Issue 9

Full text available:  pdf(208.56 KB)  html(29.84 KB) Additional Information: [full citation](#), [references](#), [citations](#), [index terms](#)

16 Markets and privacy 

Kenneth C. Laudon
September 1996 **Communications of the ACM**, Volume 39 Issue 9

Full text available:  pdf(231.63 KB) Additional Information: [full citation](#), [references](#), [citations](#), [index terms](#), [review](#)

17 Security II: Neglect of information privacy instruction: a case of educational malpractice? 

Victoria W. Romney, Gordon W. Romney
October 2004 **Proceedings of the 5th conference on Information technology education**

Full text available:  pdf(128.22 KB) Additional Information: [full citation](#), [abstract](#), [references](#), [index terms](#)

Not only should InformationTechnology (IT) Educators be knowledgeable regarding data

privacy legislation but they should be teaching correct system and database design principles to IT students in order to ensure future application design compliance with international legislative trends. Perhaps the most contentious and serious issue facing IT practitioners in the world today is data privacy. Data Privacy impacts every aspect of IT from database and application design to privacy and use policy ...

Keywords: European union directive, Gramm-Leach-Bliley, HIPAA, IT education, data privacy, database design, legal issues, legislation

18 IS '97: model curriculum and guidelines for undergraduate degree programs in information systems



Gordon B. Davis, John T. Gorgone, J. Daniel Couger, David L. Feinstein, Herbert E. Longenecker

December 1996 **ACM SIGMIS Database , Guidelines for undergraduate degree programs on Model curriculum and guidelines for undergraduate degree programs in information systems**, Volume 28 Issue 1

Full text available: [pdf\(7.24 MB\)](#) Additional Information: [full citation](#), [citations](#)

19 Special issue on persistent object systems: Orthogonally persistent object systems



Malcolm Atkinson, Ronald Morrison

July 1995 **The VLDB Journal — The International Journal on Very Large Data Bases**, Volume 4 Issue 3

Full text available: [pdf\(5.02 MB\)](#) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#)

Persistent Application Systems (PASs) are of increasing social and economic importance. They have the potential to be long-lived, concurrently accessed, and consist of large bodies of data and programs. Typical examples of PASs are CAD/CAM systems, office automation, CASE tools, software engineering environments, and patient-care support systems in hospitals. Orthogonally persistent object systems are intended to provide improved support for the design, construction, maintenance, and operation of ...

Keywords: database programming languages, orthogonal persistence, persistent application systems, persistent programming languages

20 Database privacy: balancing confidentiality, integrity and availability



Martin S. Olivier

December 2002 **ACM SIGKDD Explorations Newsletter**, Volume 4 Issue 2

Full text available: [pdf\(122.57 KB\)](#) Additional Information: [full citation](#), [abstract](#), [references](#), [citations](#), [index terms](#)

The emphasis in database privacy should fall on a balance between confidentiality, integrity and availability of personal data, rather than on confidentiality alone. This balance should not necessarily be a trade-off, but should take into account the sensitive nature of the data being stored and attempt to increase all three dimensions to the highest level possible. To achieve such a balance, technological means should be developed. The paper illustrates some of the inherent problems in database p ...

Keywords: availability, confidentiality, database privacy, dataveillance, integrity, personal data

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DIALOG(R) File 2:INSPEC
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7397595 INSPEC Abstract Number: C2002-11-7140-005
Title: Privacy rights in personal information: HIPAA and the privacy gap between fundamental privacy rights and medical information
Author(s): Davis, K.B.
Journal: John Marshall Journal of Computer & Information Law vol.19,
no.4 p.535-55
Publisher: John Marshall law School,
Publication Date: Summer 2001 Country of Publication: USA
CODEN: JCJIEI ISSN: 1078-4128
SICI: 1078-4128(200122)19:4L.535:PRPI;1-6

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Material Identity Number: C434-2002-003

Language: English Document Type: Journal Paper (JP)

Treatment: General, Review (G)

Abstract: Every technological advancement brings with it unintended consequences-some good, some not so good. With regard to health issues, developments of computer technology have impacted nearly every facet of health care. Information about a patient can be quickly accessed by physicians at nearly anytime and in nearly any location, thus providing physicians with potentially crucial information to aid in patient care. This is the good. The not so good is that the same information, and more, are equally accessible. As the amount of people with access to medical information of a sensitive nature has grown, those in the medical community and privacy advocates began to recognize the need for broad privacy protections to medical data. The result of this campaign is the Standards for Privacy of Individually Identifiable Health Information (the Privacy Rule), a set of regulations promulgated by the Secretary of Health and Human Services (HHS). The Privacy Rule was required by the Health Insurance Portability & Accountability Act of 1996 (HIPAA), then popularly known as the Kennedy-Kassenbaum Act. The Privacy Rule is important because it bridges the privacy gap between those interests deemed fundamental by the Supreme Court, and private personal information, in this case relating to medical information, that reasonable people would choose to keep out of the public domain. (182 Refs)

Subfile: C

Descriptors: data privacy; insurance data processing; legislation; medical information systems

Identifiers: privacy rights; personal information; HIPAA; medical information; Standards for Privacy of Individually Identifiable Health Information; Health Insurance Portability & Accountability Act of 1996

Class Codes: C7140 (Medical administration); C7120 (Financial computing); C6130S (Data security); C7330 (Biology and medical computing); C0230B (Legal aspects of computing)

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1/9/2 (Item 1 from file: 4)

DIALOG(R) File 4:INSPEC

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739755 INSPEC Abstract Number: C2002-11-7140-005

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File 129:PHIND(Archival) 1980-2005/Feb W1
(c) 2005 T&F Informa UK Ltd
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RATES 129 for details.
File 149:TGG Health&Wellness DB(SM) 1976-2005/Feb W1
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1/9/1 (Item 1 from file: 149)
DIALOG(R) File 149:TGG Health&Wellness DB(SM)
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02343360 SUPPLIER NUMBER: 112903437 (THIS IS THE FULL TEXT)
Cystic fibrosis adult care *: consensus conference report.
Yankaskas, James R.; Marshall, Bruce C.; Sufian, Beth; Simon, Richard H.;
David, Rodman
Chest, 125, 1, 1S(39)
Jan,
2004
PUBLICATION FORMAT: Magazine/Journal; Refereed ISSN: 0012-3692
LANGUAGE: English RECORD TYPE: Fulltext TARGET AUDIENCE: Professional
WORD COUNT: 31636 LINE COUNT: 02676

TEXT:

Key words: bronchiectasis; comprehensive health care; cystic fibrosis; nutrition; pancreatic insufficiency
Abbreviations: ADA = Americans with Disabilities Act; BMD = bone mineral density; BMI = body mass index; CBAVD = congenital bilateral absence of the vas deferens; CF = cystic fibrosis; CFF = Cystic Fibrosis Foundation; CFRD = cystic fibrosis-related diabetes; CFTR = cystic fibrosis transmembrane conductance regulator; COBRA = Comprehensive Omnibus Budget Reconciliation Act; CPT = chest physiotherapy; DEXA = dual-energy x-ray absorptiometry; DIOS = distal intestinal obstruction syndrome; DM = diabetes mellitus; ERCP = endoscopic retrograde cholangiopancreatography; FBG = fasting blood glucose; GGT = gamma-glutamyl transferase; HPOA = hypertrophic pulmonary osteoarthropathy; IBW = ideal body weight; LFT = liver function test; OGTT = oral glucose tolerance test; 25-OHD = 25-hydroxyvitamin D; PD = potential difference; PEG = polyethylene glycol; PEP = positive expiratory pressure; PFT = pulmonary function test; SSDI = Social Security Disability Insurance; SSI = Social Security Income; TOBI = aerosolized tobramycin; UDCA = ursodeoxycholic acid

No achievement highlights the striking developments of the past few decades in cystic fibrosis (CF) care more clearly than the tremendous growth of the adult CF population. This demographic shift has created the need for adult-specific CF care programs and protocols. In June 1999, the Cystic Fibrosis Foundation (CFF) convened a consensus conference to discuss the state of adult CF care. This document summarizes the findings of that meeting and incorporates information gathered since the conference convened. The recommendations embodied herein are intended to serve as a template for US adult CF programs and a resource for those wishing to find information specific to adult CF care. In cases in which prior publications sufficiently treat the subject, references are provided. In cases in which insufficient evidence was available to reach consensus, the best consensus opinions are provided. This document is not intended to replace the Clinical Practice Guidelines for Cystic Fibrosis (1) but rather to act as an adult-specific complement.

EPIDEMIOLOGY AND SURVIVAL

Of the 22,301 patients with CF in the 2000 Cystic Fibrosis Foundation Patient Registry Annual Data Report, 8,637 (38.7%) were (greater than or equal to) 18 years of age. (2) This represents a dramatic increase in the number of adults over the past 3 decades, up from about 700 (10% of all CF patients) in 1970 (Fig 1).

(FIGURE 1 OMITTED)

These changes are attributable in large part to the significant improvement in survival over the past 30 years. The median predicted survival of only 16 years in 1970 is now up to approximately 32 years. For

patients born in the 1990s, the median survival is predicted to be > 40 years. (3) The average age for all patients in the Cystic Fibrosis Foundation Registry is now > 16 years. Among adults with CF, 64% are between the ages of 18 and 29 years, 25% are between 30 and 39 years, 10% are between 40 and 49 years, and 2% are > 50 years of age. (2) The oldest living patient in the 2000 CFF Patient Registry was 78 years old. (2)

Data collection techniques and patient definition are two factors that lead some to speculate that there may be an additional 2,000 to 7,000 patients with CF in the United States who are not included in the CFF Patient Registry. Many of these patients are thought to be adults. Patients reported in the CFF Registry primarily fulfill the classic criteria for CF with both phenotypic manifestations and laboratory abnormalities consistent with CF transmembrane conductance regulator (CFTR) dysfunction (see "Diagnosis" section). If patients with CFTR dysfunction manifesting as pancreatitis, (4, 5) chronic sinusitis, (6) or congenital bilateral absence of the vas deferens (CBAVD) (7-9) were included, the number of adults with CF would be considerably higher.

The growth of the adult CF population is predicted to continue in the foreseeable future. Even without further improvements in survival over the next 5 years, the number of adults with CF in the United States in the year 2005 will likely exceed 10,000 and represent > 40% of the total CV population.

Among adults with CF, 53.9% are men, and 46.1% are women. (2) This may reflect the reported survival advantage in men. (10) The ethnic breakdown is 93.7% white, 3.2% Hispanic, 2.7% African American, and 0.4% of other ethnicity. Nearly 90% of the adults have completed high school, and more than one quarter of them have completed college. About half of the adults with CF are working full-time or part-time, and one quarter are students. Approximately one third of adults with CF are married. There were 97 live births to women with CF in 2000, representing a significant increase in births to CF patients over the past decade.

The lung function of adults with CF is highly variable. (2) Using FE(V.sub.1) as a measure, about 36% of adult patients have normal or mild lung dysfunction (ie, FE(V.sub.1), > 70% of predicted), 39% have moderate dysfunction (ie, FE(V.sub.1), 40 to 69% of predicted), and the remainder have severe dysfunction (FE(V.sub.1) < 40% predicted). The mean FE(V.sub.1) percent predicted for all adults with CF is 60.8%. As a group, adults have more severe pulmonary disease than children and are at increased risk for serious complications (Table 1).

The health-care needs of adults with CF are considerable. (2) Each year, they make an average of 4.7 CF clinic visits, experience 1.5 acute exacerbations, and are admitted to the hospital 1.0 times. In response to the increasing numbers of adults with CF, the number of adult CF programs has grown from < 10 in 1992 to > 79 in 2002.

DIAGNOSIS

Although CF is usually discovered early in life (70% by the age of 1 year), the diagnosis is being made in adults with increased frequency. Among 22,301 patients in the 2000 CFF Patient Registry, the diagnosis was established at or after the age of 18 in 881 (3.7%). (2) Patients diagnosed as adults usually present with chronic respiratory problems. As a group, they have milder lung disease, less pseudomonal infection, and are more likely to be pancreatic-sufficient than patients in whom CF is diagnosed at an earlier age. (2, 11, 12) Physicians unfamiliar with the spectrum of CF phenotypic manifestations may not consider a diagnosis of CF, thus delaying the diagnosis. Also contributing to a delay in diagnosis is the finding that some adult patients have normal or borderline sweat test results. (13-15)

Despite these differences, the criteria for establishing a CF diagnosis are the same for adults and children. (16) CF is usually suspected because of the presence of one or more typical CF phenotypic

features (Table 2). The diagnosis is confirmed by the documentation of CFTR dysfunction. When performed by an experienced laboratory in accordance with National Committee for Clinical Laboratory Standards, (17) the quantitative pilocarpine iontophoresis sweat test remains the single most useful diagnostic test for CF in adults. It should be the initial test performed in a suspected case. Although there is a spectrum of sweat chloride levels ranging from normal (< 40 mM), to borderline (40 to 60 mM), to abnormal (> 60 mM) among patients in whom CF is diagnosed during adulthood, the sweat chloride concentration will be abnormal in > 90% of those diagnosed patients. (2) However, as noted, a normal sweat chloride value cannot be used as the sole criterion for ruling out the diagnosis of CF.

In patients suspected of having CF who have a normal or borderline sweat chloride value, CFTR mutation analysis should be performed. The sensitivity of such testing is limited because current commercial panels screen for only a minority of the > 1,000 identified CF mutations, and patients who receive diagnoses after the age of 18 years are more likely to carry infrequent or unidentified mutations. (2, 11, 12) Genotype analysis was performed for 673 of the 881 patients in the 2000 CFF Patient Registry who received diagnoses at or after 18 years of age. Of these patients, two of the common (DELTAF508 mutations were identified in 142 patients (21.1%), one (DELTAF508 mutation was identified in 348 patients (51.7%), and no (DELTAF508 mutations were identified in 183 patients (27.2%). (2) For patients suspected of having CF in whom a diagnosis cannot be made on the basis of the identification of two CFTR mutations, the measurement of nasal potential difference (PD) may be used to confirm the diagnosis. (16, 18, 19) However, this technique is not available at all medical centers. More comprehensive genotype analysis by DNA sequencing is available for patients with unusual clinical and/or CFTR function tests (ie, sweat chloride or nasal PD measurements) through the CFF-sponsored Mutation Analysis Resource Center at Johns Hopkins University (Baltimore, MD; Garry Cutting, MD, Director).

Adult patients also may come to medical attention with atypical presentations such as chronic/recurrent pancreatitis, (4, 5) chronic sinusitis, (6) or CBAVD. (7-9) Men presenting with obstructive azoospermia secondary to CBAVD present a particularly challenging diagnostic problem. The majority have no other phenotypic features of CF, but 50 to 60% of those men carry one identified CF mutation, and 15 to 20% are compound heterozygotes. A diagnosis of CF should be assigned to such patients only if there is documentation of elevated sweat chloride values, two CF mutations, or an abnormal nasal PD measurement. (16) It is important to consider alternative diagnoses (eg, immunodeficiency, ciliary dyskinesia, and Young syndrome) in patients with atypical presentations.

People who receive diagnoses of CF as adults may be overwhelmed by the implications of a disease that leads to premature death for many children and young adults. It is important for the CF care team to educate such patients about the disease. In particular, they should be informed that patients who receive diagnoses in adulthood often have much better prognoses than patients who receive diagnoses during early childhood.

STANDARD CARE

Overview

Data comparing the relative effectiveness of various approaches to adult CF care (ie, multidisciplinary vs subspecialty vs primary care-based) are lacking. However, based on the strong association between the establishment of comprehensive CF Care Centers and improved patient outcomes, the committee strongly recommends a multidisciplinary approach modeled on the highly successful pediatric CF care system. The health-care team should include at least a part-time commitment from a physician, nurse, respiratory therapist, dietitian, and social worker. Ideally, 'all members of the team should have specific training in adult CF care. Back-up personnel should be available in the event that a team member is unable to

perform his or her duties. The pulmonary and GI/nutritional manifestations of the disease predominate in adults as in children, but several other issues also emerge.

Comprehensive Care

The primary objectives of the adult health-care team are to: (1) ensure optimum care; (2) facilitate access to pertinent medical resources; (3) coordinate care among specialists and primary care practitioners; and (4) support quality of life and independence for each patient. Frequent patient contact with the Center is necessary to accomplish these objectives. In general, quarterly visits are sufficient, although some patients with special needs or advanced disease may require more frequent attention. The Adult CF Care Team may function in primary care capacity or in concert with an independent primary care practitioner. Coordination and communication with other medical professionals involved in the patient's care are essential.

The optimal management of CF requires input from all members of the health-care team. Evaluation and intervention by team members should be individualized to suit each patient's circumstances. However, a minimum of one comprehensive evaluation per year by each team member (ie, nurse, respiratory therapist, dietitian, and social worker) is recommended. These evaluations should encompass an assessment of adherence with therapies and the identification of relevant psychosocial issues as well as specific medical issues. When the center is serving in a primary care capacity, health maintenance (eg, vaccinations and cancer screening) should be provided according to national guidelines for age and gender. Ideally, the center should have a case management conference or other mechanism in place for a periodic review of the status of each patient and the formulation of a treatment plan. These assessments should be documented in the medical record and communicated with other health-care professionals involved in the care of the patient.

Pulmonary Disease

Assessment: The pulmonary status of patients should be regularly monitored by an assessment of symptoms, a physical examination, and, on most visits, spirometry. FE(V.sub.1), expressed as the percent predicted of a healthy nonsmoking reference population, is accepted as the single most useful objective measure of pulmonary status. (20) Oxygen saturation should be measured routinely in patients with moderate-to-severe pulmonary disease to assess the need for supplemental oxygen. The measurement of oxygen saturation during exercise and/or sleep may be indicated in some situations.

A complete microbiological assessment of expectorated sputum, including antibiotic susceptibility testing, should be performed at least on an annual basis, and preferably on a quarterly basis. Oropharyngeal swab cultures, (21, 22) which are commonly obtained from children who do not produce sputum, have not been fully studied in adults. The microbiology laboratory should follow published guidelines (23) for the processing of CF sputum in order to isolate the wide range of organisms found in these specimens. Multiply-resistant Gram-negative organisms, such as *Burkholderia cepacia*, *Stenotrophomonas maltophilia*, and *Achromobacter xylosoxidans*, are found in up to one third of adults with CF. (24) This has significant implications for disease management and infection control. The microbiology laboratory must be capable of distinguishing among these organisms and performing extended antimicrobial susceptibility panels if necessary. Antibiotic synergy testing may be helpful in some situations and is available at the CFF-sponsored reference laboratory at Columbia University for patients who have received follow-up at accredited CF care centers. (25) Confirmed or suspected *B cepacia* isolates should be sent to the CFF-sponsored reference laboratory at the University of Michigan (26) for the confirmation of identity and further characterization.

The role of chest imaging in the monitoring of adults with CF has not

been extensively studied. Standardized chest radiograph scoring may be useful in documenting the progression of disease or response to therapeutic intervention. (27, 28) The committee did not support the use of annual surveillance chest radiographs but reached a consensus opinion that posterior/anterior and lateral chest films should be obtained every 2 to 4 years in patients with stable clinical status. Imaging also should be considered for patients with signs or symptoms consistent with a significant acute pulmonary exacerbation, pneumothorax, lobar atelectasis, or hemoptysis. Chest CT scans may be appropriate in certain clinical situations but cannot be recommended on a routine basis.

Additional diagnoses such as asthma, nontuberculous mycobacterial infection, allergic bronchopulmonary aspergillosis, sinus disease, and gastroesophageal reflux, should be considered in patients whose symptoms, clinical course, or response to treatment are atypical for CF.

Treatment: The cornerstones of treatment for those with CF are antibiotic therapy, airway clearance, and nutritional support; which are similar for children and adults. The reader is referred to several reviews (29-32) for a more detailed discussion of the specific components of a standard treatment regimen, which include antibiotic therapy for pulmonary exacerbations, and chronic suppressive therapy, airway clearance and exercise, therapy with mucolytic agents, bronchodilators, and anti-inflammatory agents, supplemental oxygen, and nutritional support. Because of the potential complexity, all aspects of the medical regimen should be reviewed on a regular basis with an assessment of adherence and potential side effects from medications.

Basic principles in the treatment of pulmonary exacerbations will be covered first, followed by a discussion of the various components of maintenance therapy for the treatment of pulmonary disease. Nutrition will be covered later in the article.

PULMONARY EXACERBATIONS: Pulmonary exacerbations are common in adults with CF. The approach to treatment of an exacerbation described in Clinical Practice Guidelines for Cystic Fibrosis (1) of the CFF is applicable to the adult population. Specific antibiotics are selected on the basis of a recent sputum culture. *Pseudomonas aeruginosa* is by far the most common pathogen found in adults with CF. Therapy with fluoroquinolones is often used for mild-to-moderate exacerbations. Two antipseudomonal antibiotics are used in combination (eg, a (beta)-lactam and an aminoglycoside) for the treatment of moderate-to-severe pulmonary exacerbations. Clinicians must be aware that for many antibiotics, differences in the volume of distribution and the rate of elimination in CF patients require higher doses and shorter dosing intervals. (33)

As noted above, adults are more likely¹ to be infected with multidrug-resistant organisms such as *B* *cepacia*. Antibiotic combinations are typically used for the treatment of exacerbations related to these organisms. Some centers treat empirically and others use synergy testing to select a treatment regimen. Inhaled antibiotics are at times used in combination with parenteral agents. The optimal approach has not been validated in clinical trials. In order to prevent person-to-person spread of these organisms, (34) the center must have rigorous infection control practices in place in the outpatient clinics and the inpatient units.

CHRONIC SUPPRESSIVE ANTIBIOTIC THERAPY: Chronic suppressive antibiotic therapy often is employed because the treatment of pulmonary exacerbations will not eradicate the lung infection. Aerosolized tobramycin (TOBI) has been the most thoroughly studied chronic suppressive therapy. In two large, multicenter, double-blind, placebo-controlled trials conducted over a 24-week period, treatment with TOBI was found to produce significant improvement in pulmonary function, to decrease the density of *P* *aeruginosa* in sputum, and to decrease the number of days that subjects were hospitalized. (35) These studies included patients with moderate-to-severe pulmonary disease, which was defined as an FE(V._{sub}1) between 25% and 75%

of predicted. Subset analyses demonstrated that adolescents had the greatest response, although all age groups and disease severity categories showed significant improvement from the therapy. A 24-month open-label follow-up (36) of these trials demonstrated sustained improvement in FE(V.sub.1) compared to the group that had initially received placebo.

A significant long-term concern in using chronic suppressive therapy of any type is the emergence of antimicrobial resistance. The TOBI trials showed no increase in the prevalence of *B. cepacia* or other resistant organisms in the TOBI-treated group. There was a modest but detectable shift in the minimum inhibitory concentrations of the *P. aeruginosa* strains infecting the TOBI-treated subjects. The sustained improvement in pulmonary function appears to outweigh the risk of tobramycin resistance that may develop over time, but this must be carefully considered for each individual.

Who should be considered for this therapy? Any adult patient chronically infected with *P. aeruginosa* is a potential candidate. Certainly, any patient who falls within the patient selection criteria for the phase III clinical trial (35) (ie, those with FE(V.sub.1) between 25% and 75% of predicted) deserves serious consideration for inclusion in a therapeutic trial of this drug. More severely affected patients also may benefit, but they should be carefully monitored during the initiation of therapy. The more difficult issue is the case of the mildly affected patient with an FE(V.sub.1) of > 75% predicted. Interventions at this point in the disease course may have a profound impact on the subsequent course of disease. The current trend is toward more aggressive therapy and earlier intervention. TOBI should be considered for this mildly affected group, particularly for those in whom the pulmonary disease is more active. Indications of more active disease may include increased symptoms, declining pulmonary function, or increased frequency of pulmonary exacerbations. All ongoing clinical trial of TOBI in this mildly affected patient population hopefully will shed additional light on this issue.

One caveat with this therapy relates to the delivery of the antibiotic to the airways. Particle size is critical to antibiotic deposition. Too small a particle will tend to deposit in the distal air spaces and alveoli where significant systemic absorption may occur. Too large a particle will tend to deposit in the central airways. TOBI should be administered with the nebulizer systems that were validated in the clinical trials. (35)

The data presented above specifically refer to the TOBI preparation of tobramycin. A number of smaller, less rigorous clinical trials of other formulations of tobramycin and gentamicin using a variety of dosing regimens ranging from 80 to 600 mg, two to three times a day have been reported. (37-42) Because of methodological concerns with these trials, the efficacy and safety of preparations other than TOBI have not been established.

Inhaled colistin (Coly-Mycin; Monarch Pharmaceuticals, Bristol, TN) has been reported to be of benefit in case series and uncontrolled clinical trials. (43, 44) The rarity of antimicrobial resistance has been touted as an advantage with this drug; however, the transmission of colistin-resistant *P. aeruginosa* recently has been reported. (45) Bioavailability from the aerosolized route of administration has not been studied adequately. Safety is also a concern due to the fact that bronchospasm occurs in a substantial proportion of patients after the nebulization of this medication. (45, 46) In a recent short-term comparison trial (47) in the United Kingdom, pulmonary function improved in the TOBI-treated group, but there was no significant change in the colistin group. Thus, the efficacy and safety of aerosolized colistin has not been established.

Scheduled parenteral antipseudomonal therapy is an alternative suppressive strategy, that has been popular in Denmark. Retrospective and

uncontrolled reports have suggested improved survival in patients treated with 2-week courses of therapy, four times per year irrespective of symptoms. (48) However, a randomized trial (49) comparing suppressive IV antibiotic therapy administered four times per year vs standard treatment for symptoms of an exacerbation showed no difference in outcomes between the two groups. This approach cannot be recommended at this time.

There are few data and no convincing evidence to support the use of chronic oral antibiotic therapy in the adult CF population. However, therapy with macrolide antibiotics have garnered attention based on their effectiveness in the treatment of diffuse panbronchiolitis. (50) Promising preliminary observations suggested a clinical benefit in CF (51-53) and led to several randomized, controlled clinical trials. A double-blind, placebo-controlled azithromycin trial in the United Kingdom (54) included 41 children, 20 of whom did not have persistent *P aeruginosa* infection. The crossover study design included two 6-month treatment phases and an intervening 2-month washout period. The median relative improvement in FE(V.sub.1) in the azithromycin phase was 5.4%. Oral antibiotic use was reduced, but the number of pulmonary exacerbations and courses of IV antibiotics did not differ in the azithromycin and placebo phases. A 3-month randomized, placebo-controlled, double-blind trial of azithromycin enrolled 60 stable adults in Australia who were chronically infected with *P aeruginosa*. (55) By chance, the treatment assignments resulted in significant differences between the azithromycin and placebo groups. At baseline, the placebo group contained more men, and on average the patients were taller, heavier, and had better lung function than those in the azithromycin group, requiring adjustments in their statistical analyses. Nevertheless, the azithromycin group had a 3.6% relative improvement in FE(V.sub.1), had undergone fewer courses of IV antibiotic therapy, and had fewer hospital days. A 24-week, multicenter, randomized, placebo-controlled, double-blind trial of azithromycin (56) in the United States enrolled 185 patients who were (greater than or equal to) 6 years of age and were chronically infected with *P aeruginosa*. The azithromycin group experienced a 6.2% treatment benefit in relative FE(V.sub.1) percent predicted change from baseline, decreased pulmonary exacerbations, decreased hospitalizations, and a 0.7-kg weight gain compared to the placebo group. The drug was well tolerated in all three trials. In summary, despite differences in patient populations, study design, and treatment regimens, all three trials demonstrated clinical improvement with azithromycin therapy. The mechanism of action remains to be elucidated. Based on this evidence, we believe that azithromycin should be considered for CF patients who are (greater than or equal to) 6 years of age who are chronically infected with *P aeruginosa*. The US trial regimen was 500 mg thrice weekly for patients weighing (greater than or equal to) 40 kg, and 250 mg thrice weekly for patients weighing 25 to 40 kg. A sputum smear and culture for acid-fast bacilli should be obtained prior to initiating chronic macrolide therapy because of the slight risk of having an undiagnosed nontuberculous mycobacterial infection developing macrolide resistance. For patients receiving chronic macrolide therapy, a smear and culture for acid-fast bacilli should be obtained every 6 months.

AIRWAY CLEARANCE AND EXERCISE: There are a variety of airway clearance techniques available. Conventional chest physiotherapy (CPT) is the technique of percussion and postural drainage. Despite the absence of randomized, controlled clinical trials, the available evidence and clinical experience supporting conventional CPT appears to be convincing. (57, 58) Potential problems with this modality include hypoxia, particularly in patients with severe lung disease, (59) and gastroesophageal reflux. (60) Furthermore, CPT is physically demanding and time-consuming for both the patient and Iris or her support person(s). Poor adherence is common. (61)

As patients become older and more independent, they frequently seek airway clearance methods that can be performed without assistance. Several

alternative modalities have been developed, including active cycle breathing, forced expiratory technique, positive expiratory pressure (PEP) devices, autogenic drainage, and high-frequency chest wall oscillation systems (ie, The Vest, formerly called ThAIRapy Vest (Advanced Respiratory; St. Paul, MN); the Flutter device (Axcan Scandipharm; Birmingham, AL); and the intrapulmonary percussive device). A detailed description of each of these methods can be found in a review. (62) Meta-analysis suggests that CPT resulted in greater sputum production than no treatment, and that the addition of exercise improved FE(V.sub.1). No other differences between modalities were found. (63)

Expiratory resistance or PEP devices may promote mucus clearance by preventing airway closure and increasing collateral ventilation. They are used via a mask or a mouthpiece and can be adapted for the concomitant delivery of bronchodilators. These devices have been extensively studied in Europe, with most trials demonstrating equivalence to conventional CPT. (64) A long-term (ie, 1 year) trial showed that PEP therapy was superior to conventional CPT with respect to maintaining pulmonary function. (65) A report (66) demonstrated that PEP was also more effective than the Flutter device in maintaining pulmonary function over the course of a 1-year trial. The devices are relatively inexpensive, portable, and well-tolerated, although some patients find them fatiguing. Theoretical concern that they might increase the risk of pneumothorax has not been borne out in practice.

The Vest is a chest wall compression and oscillation system composed of a fitted vest coupled to a pneumatic compressor. Therapy is delivered to the entire chest at the same time with the patient in a seated position. This allows the administration of nebulized medications during the therapy session, minimizing the patient's time commitment; but the major advantage over conventional CPT is the degree of independence afforded to the patient. A prospective study (67) demonstrated that use of The Vest is equivalent to conventional CPT in patients hospitalized for a pulmonary exacerbation. The major disadvantages with this system are the expense and lack of portability. Some severely affected patients complain that it is harder to breathe during the treatment. Others, particularly those with indwelling venous access devices (eg, the Port-a-Cath; Deltec, Inc; St. Paul, MN), may find The Vest uncomfortable.

Additional clinical trials are needed to define the optimal airway clearance regimens. To provide meaningful results, such studies must involve an adequate number of patients observed over a time frame of at least several months. Sputum production has been the primary outcome variable in many of the published trials to date, but it may prove to be an unreliable marker of efficacy. The preservation of pulmonary function provides more convincing evidence of efficacy.

Physical activity augments airway clearances (68-70) and should be viewed as an important adjunct to the airway clearance techniques described above. A randomized clinical trial (71) demonstrated that regular aerobic exercise attenuates the decline in pulmonary function over a 3-year period compared to a control group. In addition, appropriate vigorous physical exercise enhances cardiovascular fitness, increases functional capacity, and improves quality of life. The level of physical fitness, as measured by maximal oxygen uptake, correlates with survival in CF. (72) For these reasons, with the exception of those patients whose clinical condition prevents it, all adults with CF should be encouraged to exercise.

Ideally, patients should learn exercise techniques under the supervision of a qualified physical therapist. In patients with moderate-to-severe pulmonary disease, it is important to ensure adequate oxygenation during exercise. (73) Aerobic activities, such as swimming, jogging, and cycling, are the most commonly recommended forms of exercise. Patients should be encouraged to exercise several times per week. Pulmonary rehabilitation regimens previously targeted for adults with emphysema and chronic bronchitis will likely prove to be effective in the CF population.

The health-care team must assist the patient and family in tailoring an airway clearance regimen that provides the best fit to the patient's lifestyle and activities. Typically, this consists of conventional CPT and/or a suitable alternative airway clearance technique, combined with an aerobic exercise program. The frequency and duration of each treatment should be individualized. Patients with minimal-to-mild symptoms may only require one session a day, whereas others with a greater volume of thick secretions may need three or more sessions per day. Developing an individualized regimen that is acceptable to the patient and the physician is a trial-and-error process that requires staff who are well-versed in airway clearance techniques and exercise physiology. Care providers must consistently encourage and monitor adherence, a major issue with this aspect of care, and modify the regimen as necessary.

MUCOLYTIC AGENTS: Recombinant human DNase (also known as (alpha)-dornase or Pulmozyme; Genentech, South San Francisco, CA) decreases the viscosity of CF sputum by catalyzing extracellular DNA into smaller fragments. (74) A large phase III randomized, double-blind, placebo-controlled trial showed a modest improvement in pulmonary function in the DNase-treated groups (5.8% and 5.6% relative improvement in FE(V.sub.1) from baseline, respectively, in the groups treated once and twice a day compared to the placebo group), decreased pulmonary exacerbation rate (28% and 37% reductions, respectively, in the age-adjusted risk of pulmonary exacerbations in patients treated once and twice a day compared to the placebo group), and some improvement in CF-related symptoms. (75) The decrease in respiratory tract infectious resulted in fewer days in the hospital and fewer days receiving parenteral antibiotics for the DNase-treated groups.

It is not clear whether the differences in pulmonary function and respiratory tract infection rate observed in this clinical study will impact mortality. The majority of participants in the phase III trial continued receiving DNase for up to 2 years in an open-label extension of the study. Age-adjusted and height-adjusted pulmonary function at the 2-year point was declining at the same rate as in the placebo treated group during the randomized portion of the trial. (76) This would suggest that DNase had perhaps delayed but not prevented progression of the disease. However, it should be kept in mind that this trial consisted of a patient population that was not representative of the CF population as a whole. Specifically, this was an older group of patients with more significant obstructive airways disease. Another randomized trial (77) in even more severely affected patients with FVC values of < 40% also showed improvement in FE(V.sub.1) from baseline (DNase-treated group, 9.4%; control group, 2.1%) over the 12-week study period.

More recently, the results of the Pulmozyme Early Intervention Trial have been published. (78) This phase III, double-blind, placebo-controlled trial in children with mild disease showed modest improvement in pulmonary function and a reduction in pulmonary exacerbations over a 2-year period. The long-term impact of treating a mildly affected population of patients is not blown at present.

Postmarketing clinical experience has confirmed the relative safety of DNase. Hoarseness, voice alteration, and pharyngitis are the major adverse events related to DNase, and, in most cases, these symptoms are self-limited and do not require the cessation of drug therapy. (75) In addition, concerns that DNase might release neutrophil elastase bound to DNA and thereby exacerbate the inflammatory state have been resolved. Shah et al (79) found a decline in neutrophil elastase activity and interleukin-8 levels in the sputum of CF patients treated with DNase over a 6-month time period.

Should DNase be prescribed to all CF patients? Patients with chronic productive cough, particularly those with moderate-to-severe obstructive airways disease, should be considered for a therapeutic trial of

once-per-day DNase for a period of several months. The drug can be started safely during an acute pulmonary exacerbation (80) as well as during a stable period. Patients should be monitored by symptoms, pulmonary function tests (PFTs), and pattern of exacerbations. Unfortunately, there are no clear-cut criteria to judge clinical response. In the phase III trial, subjects who did not show an improvement in spirometry still had a reduction in pulmonary exacerbation rate. (75) Often the patient's subjective response to the drug is a major factor in the decision to continue therapy for the long term. This is certainly reasonable; however, patients should be encouraged to give the drug a fair trial of at least several months. Some severely affected patients seem to benefit symptomatically from a twice-a-day regimen.

Mildly affected patients also should be made aware of this drug and its potential benefits. The relatively high cost of DNase may factor into a decision about whether to prescribe this therapy for a mildly affected adult, but a therapeutic trial in an individual patient is justifiable and should be considered with currently available information.

There are no well-validated alternative mucolytic agents available at this time. N-acetylcysteine reduces viscosity of sputum in vitro, presumably by breaking disulfide bonds. The nebulized form of the drug has been used in CF patients but has not been carefully studied. Trials (81, 82) in patients with COPD have not demonstrated a significant beneficial effect. Furthermore, the drug can be very irritating to the upper airway and can cause bronchoconstriction. (83) Some European studies have suggested a modest benefit from oral N-acetylcysteine, particularly in patients with moderate-to-severe disease, (84) but others have not confirmed these findings. (85) It is not even clear that adequate amounts of orally administered drug penetrate into the airways to have a mucolytic effect. (86) For these reasons, the efficacy and safety of N-acetylcysteine has not been established.

There has been renewed interest in the use of nebulized hypertonic saline solution to facilitate airway clearance. It improves mucociliary clearance, (87) likely by its effects on sputum viscoelasticity. (88, 89) A short-term (ie, 2-week) clinical trial (90) demonstrated that nebulization of a 6% saline solution twice a day resulted in an improvement in PFTs compared to a control group that nebulized an isotonic saline solution (15.0% vs 2.8% improvement, respectively, in FE(V.sub.1) from baseline; p = 0.004). Hypertonic saline solution has the potential to cause bronchospasm in patients with CF, (91) but this may be preventable by pretreatment with a bronchodilator. The preliminary data are promising, but it is premature to recommend the widespread use of hypertonic saline solution at this time.

BRONCHODILATORS: The majority of patients with CF demonstrate bronchial hyperreactivity at least some of the time. (92) Bronchodilators have therefore become a standard component of the therapeutic regimen. Nebulized (beta)-adrenergic agonists are the most commonly prescribed agents. They are often used to provide symptomatic relief and, prior to CPT, to facilitate clearance of the airways. Konig and colleagues (93) reported that maintenance albuterol treatment reverses the progressive downhill course in lung function in CF patients. A longer term, placebo-controlled, double-blind study (94) also showed sustained improvement in PFT scores in a group of patients treated with albuterol, but the difference from the control group was not statistically significant, likely because of all insufficient number of study subjects.

These agents are, in general, well-tolerated in the CF population. Most patients demonstrate improved pulmonary function with bronchodilators, (95, 96) but the occasional patient may actually worsen with bronchodilator therapy. (97) Airflow may decrease paradoxically, or hyperinflation may worsen because of smooth muscle relaxation and decreased airway elasticity. Periodic pulmonary function testing and careful attention to symptoms will identify those few patients in whom bronchodilator therapy is

counterproductive.

In summary, the potential benefits of inhaled (beta)-agonist agents outweigh the risks. They should be considered for all adults with CF. Long-acting aerosolized (beta)-adrenergic agonists also may have a role in this disease. Salmeterol has been associated with better preservation of pulmonary function and oxygenation through the night. (98, 99) In addition, a 24-week randomized, double-blind, placebo-controlled crossover study (100) involving 23 patients with mild-to-moderate pulmonary disease showed that high-dose salmeterol (100 (micro)g/d) was equally safe, and was associated with better pulmonary function, fewer antibiotic interventions, and fewer respiratory symptoms compared to twice daily therapy with nebulized albuterol. Oral preparations have no advantage over the inhaled medications in reversing bronchospasm, (101) so they are not commonly used.

Anticholinergic bronchodilators may be helpful for some patients with CF. Atropine has been associated with unacceptable systemic side effects, but ipratropium bromide is very poorly absorbed and much better tolerated. It has been shown to have some benefit in asthma (102) and may be of use in CF patients as well. Weintraub and Eschenbacher (103) observed that ipratropium may be more effective than (beta)-adrenergic agonists in adults with CF. Adults may have less bronchospasm but more secretions than children. The airway of the adult CF patient may more closely mimic that of the adult with chronic bronchitis, and therefore may be more responsive to the effects of a parasympathomimetic agent. Some patients appear to benefit from combination therapy with a (beta)-adrenergic agonist and an anticholinergic agent. (104) A therapeutic trial of combination therapy is indicated for patients with bronchospasm that is not well-controlled by (beta)-agonist therapy alone.

Theophylline increases mucociliary clearance, diaphragmatic contractility, and CNS respiratory drive. (105) Unfortunately, it has a narrow therapeutic range that requires the monitoring of plasma concentrations. In addition, a variety of adverse effects occur with this drug, including nausea, vomiting, and gastroesophageal reflux, which limits its utility in patients with CF.

ANTI-INFLAMMATORY THERAPIES: Some patients with CF have asthma or asthma-like symptoms that require more than therapy with bronchodilators alone. The full asthma armamentarium can be used to treat their bronchospasm. Inhaled or oral glucocorticoids seem to be generally more efficacious than cromolyn or nedocromil, but both classes of drugs are widely used in the treatment of CF patients. Other patients with CF require glucocorticoids for the treatment of allergic bronchopulmonary aspergillosis. We will not focus on these issues but rather will address the role of anti-inflammatory agents for the nonasthmatic patient with CF with chronic airways infection and inflammation.

Short-term therapy (3 weeks) with daily corticosteroids in stable patients with severe obstruction showed no benefit. (106) A population with less severe disease treated with 2 weeks of daily therapy with corticosteroids (2 mg/kg/d), followed by alternate-day steroid therapy for an additional 10 weeks (1 mg/kg every other day), showed improvement in pulmonary function, and a decrease in serum cytokine and IgG levels. (107) A longer study (108) (4 years) of therapy with alternate-day steroids (2 mg/kg every other day) also suggested a benefit from steroids with respect to pulmonary and nutritional parameters. This promising result led to a larger multicenter randomized trial comparing alternate-day therapy with prednisone at 2 mg/kg and 1 mg/kg to placebo. This trial enrolled only children and adolescents with CF, but the results are of interest to adult care providers. The higher dose group was discontinued because of an unexpectedly high incidence of cataracts, glucose intolerance, and growth retardation. (109) The 1 mg/kg and placebo groups continued to the end of the 4-year trial. The steroid-treated group showed benefit with respect to pulmonary function, particularly the subset of patients colonized with P

aeruginosa. (110) However, that benefit was at the expense of growth. A subsequent analysis (111) showed that steroid-treated men had persistent growth impairment after steroid therapy was discontinued, as indicated by reduced adult height in comparison with the placebo group. Bone density was not an end point in this trial but is another significant concern with this therapy. In summary, the data suggest that corticosteroids may have a beneficial effect but at significant cost. For this reason, long-term oral corticosteroid therapy, even in an alternate-day regimen, probably should be avoided if possible. These studies do, however, suggest that an anti-inflammatory approach has promise.

Therapy with inhaled steroids is a potential way to reduce inflammation without significant systemic adverse effects. Relatively low doses of inhaled beclomethasone (400 (micro)g/d) showed no effect on various markers of airways inflammation. (12) Higher doses of inhaled steroids have shown some promise in preliminary studies, (113, 114) but larger trials with longer term data are needed before this therapy can be recommended.

High-dose ibuprofen therapy (20 to 30 mg/kg, up to 1600 rag, bid) slowed the progression of pulmonary disease in mildly affected patients (ie, FE(V.sub.1) > 60% of predicted), particularly in children 5 to 12 years of age. (115) It is important to emphasize that a pharmacokinetic study should be done to verify that therapeutic blood levels of the drug have been achieved. Close monitoring for adverse events is also important, including a semiannual check on renal function. The potential risks of ibuprofen (GI and renal) should be carefully weighed in deciding whether to treat mildly affected adults. There are no data to support this therapy in patients with moderate-to-severe obstructive airways disease (ie, FE(V.sub.1) < 60% of predicted). Because of concern about an increased risk of hemoptysis, high-dose ibuprofen therapy should be avoided in this subset of patients.

The use of leukotriene modifiers in the CF population deserves careful study. These drugs have some attractive features, but, given the scarcity of data in CF patients, they cannot be recommended at this time.

OXYGEN: Clinically apparent cor pulmonale is a poor prognostic indicator. (116) The goals should be to prevent the development and/or progression of pulmonary hypertension. Data from Toronto (117) demonstrate that subclinical pulmonary hypertension develops in a significant proportion of patients with CF and is strongly correlated with hypoxemia, independent of pulmonary function. Furthermore, subclinical pulmonary hypertension appeared to be associated with increased mortality compared to a group with a similar degree of spirometric impairment without pulmonary hypertension.

The most important therapy for the prevention of pulmonary hypertension is supplemental oxygen, but there are limited data available on its use in treating CF. Zinman et al (118) were unable to demonstrate a beneficial effect of nocturnal oxygen therapy in patients with CF. However, there were several weaknesses in this study. A relatively small number of patients were enrolled, and some of them did not meet the usual criteria for receiving supplemental oxygen therapy. In addition, oxygen was used only at night for an average of 7.0 (+ or -) 1.9 h. In the absence of persuasive data in CF patients, we must turn to the literature on oxygen administration in COPD patients.

Supplemental oxygen has been shown to improve exercise tolerance and survival in COPD patients. The Nocturnal Oxygen Therapy Trial Group (119) and the Medical Research Council Working Party (120) both demonstrated that oxygen administration improved survival in severely affected, hypoxic COPD patients. The criteria for supplemental oxygen therapy in these studies (ie, Pa(O₂).sub.2), < 55 mm Hg during the daytime while breathing room air or < 59 mm Hg in the presence of pedal edema, polycythemia, or ECG evidence of impairment of the right side of the heart) have been widely adopted by

the medical community. Continuous oxygen therapy is indicated for such patients.

Patients are more likely initially to develop hypoxemia with exercise or during sleep. (121-123) Clinicians must be aware of this and must intermittently screen patients with moderate-to-severe pulmonary disease accordingly. Oxygen is indicated during exercise if the exercise oxygen saturation level falls below 88 to 90%. Nocturnal oxygen therapy is indicated if oxygen saturation is < 88% to 90% for (greater than or equal to) 10% of the total sleep time.

Complications: In general, adults with CF have more severe pulmonary disease than children. This puts them at higher risk for serious complications such as pneumothorax and massive hemoptysis (Table 1). The adult care team must have expertise in dealing with these medical emergencies in a timely and proficient fashion. The CFF consensus statement on pulmonary complications (124) details an approach to these problems.

The majority of CF patients die in adulthood of respiratory failure. The health-care team must be prepared to deal with the complex medical and psychosocial end-of-life issues (see "End-of-Life Options" section). The issue of advanced care directives should be addressed in the clinic with all patients, particularly in severely affected patients, when their conditions are stable. An individual patient's decisions about end-of-life issues and lung transplantation may impact the health-care team's treatment approach, including decisions about ICU admission, ventilatory support, and management of pneumothorax.

Exocrine Pancreas

Diagnosis of Pancreatic Insufficiency: Eighty-five to 90% of patients with CF have exocrine pancreatic insufficiency, which is defined as elevated fecal fat excretion. (125) The majority of adults with CF have exocrine pancreatic insufficiency, although those with mild mutations of CFTR may have residual pancreatic function and may not require supplemental pancreatic enzymes. These patients are, however, at increased risk for acute or recurrent episodes of pancreatitis. (4, 5) The decision to treat a patient with enzyme supplements rests on demonstrating the presence of steatorrhea. This generally correlates with symptoms of diarrhea, foul-smelling greasy stools, weight loss or poor weight gain, flatus and abdominal discomfort, and fat-soluble vitamin deficiency. For young adults who received diagnoses during childhood, enzyme supplementation should be continued. For newly diagnosed adults, a 72-h fecal fat collection should be performed while the patient is on a fixed oral fat intake or with dietary records. Fecal fat excretion ((grams of fat excreted/ grams of fat ingested) x 100%) can be calculated from these data. Fecal fat excretion of > 7% indicates steatorrhea in all adult and mandates the initiation of pancreatic enzyme and vitamin supplementation (see "Nutrition" subsection). In specialized CF centers, the quantitative assessment of pancreatic excretory function may be performed to better define the stimulated output of pancreatic enzymes into the duodenum, (126) but this is not necessary on a routine clinical basis. Levels of fecal chymotrypsin, (127) serum pancreatic trypsinogen, (128) and fecal pancreatic elastase-1 (129) may be low in patients with pancreatic insufficiency, however, these tests have not been fully evaluated for use in adults with CF. Recent evidence (130) has suggested that the fecal pancreatic elastase 1 immunoassay may prove to be a noninvasive, simple, and reproducible method of assessing pancreatic function.

Pancreatic Enzyme Supplements: Most modern pancreatic enzyme products are capsules that contain enteric-coated microencapsulated enzymes, either as microspheres or microtablets. The enteric coating prevents inactivation of the enzymes in the acidic gastric environment. The dissolution of generic microspheres or microtablets may not be equivalent to that of proprietary brands. (131) The substitution of one brand for another by the pharmacist may result in vastly different clinical responses despite equal

enzyme doses and should not be allowed. The ratio of proteases to lipases differs in various brands of enzymes; however, it is uncertain whether this is clinically relevant. The US Pharmacopoeia requirements state that enzyme products may contain not < 90% of the amount stated on the label but do not set an upper limit for the content. The manufacturers usually overfill the capsules to compensate for enzyme degradation during storage. (132)

Enzyme supplements should be given with meals and snacks, with the number of capsules divided between the beginning and the end of the meal. Some patients may be able to take all of the capsules at the beginning of the meal without problems. Enzyme dosing can be calculated on the basis of the amount of fat ingested with each meal. (133) In general, patients need 500 to 4000 U lipase per gram of fat ingested per day. This method of dosing mimics the body's response to adjusting pancreatic enzyme excretion but may be tedious to calculate. Although less physiologic, it is frequently more practical and convenient to determine the enzyme dose based on body weight. (134) Adults should start with approximately 500 U lipase per kg body weight per meal, and half of that with snacks. If this dose quickly corrects the fat malabsorption, then attempts should be made to reduce the dose to the minimum effective dose. If symptoms of steatorrhea continue (or if the results of 72-h fecal fat collection tests are still abnormal), then the dose should be increased in increments of approximately 150 to 250 U lipase per kilogram per meal until symptoms improve, up to a maximum of 2500 U lipase per kilogram per meal (or 4000 U lipase per gram of fat per day). Doses higher than this level should be used with caution because of the risk of the occurrence of fibrosing colonopathy with higher doses. (134-136) Patients receiving doses > 2,500 U lipase per kilogram per meal should be reevaluated, and attempts should be made to reduce the dose of enzyme supplements.

Poorly Responding Patients: Some patients may continue to have symptoms of steatorrhea despite taking appropriate doses of enzyme supplements. Adherence to the enzyme therapy should be assessed in this situation. Giving some of the enzyme capsules at midmeal may be of some benefit. The hyperacidity of the upper GI tract in CF patients is one of the most common reasons for suboptimal response to enzyme therapy. Gastric acid output may not be neutralized because of inadequate bicarbonate secreted by the pancreas. This retards the dissolution of the enteric-coated microspheres or microtablets. (137) Drugs that reduce gastric acid production (eg, (H.₂.sub.2)-blockers and proton-pump inhibitors) may help to improve the dissolution of these products and reduce steatorrhea. (138, 139) Other therapeutic considerations include the addition of non-enteric-coated, powdered preparations (eg, Viokase; Axcan Scandipharm), or a trial of an alternate brand of microencapsulated product with a different dissolution profile.

Short bowel syndrome, previous intestinal resections, and rapid GI transit are other conditions in which the microencapsulated enzyme preparations may not dissolve well. The addition of small amounts of pancreatic enzyme non-enteric-coated, powdered preparations with or without gastric acid suppression may improve fat digestion in the more proximal intestine and may improve steatorrhea. In patients who have continued symptoms despite adequate enzyme supplementation and acid suppression, other diagnoses should be considered, including infectious gastroenteritis, parasitic infestation (eg, giardiasis), lactose intolerance, bacterial overgrowth of the small intestine, cholestasis, Clostridium difficile disease, celiac disease, short bowel syndrome, Crohn disease, food allergies, or intestinal tumors. Appropriate evaluation and treatment for these disorders should be conducted when indicated. (140)

Nutrition

General Principles: The importance of nutritional status in the long-term survival of patients with CF is well-documented. (141-144) The prevention of malnutrition should be a primary goal of the health-care

team. A standard North American diet with 35 to 40% of calories from fat is recommended. (133) The tendency to restrict fat consumption in these patients should be discouraged since dietary fat is the highest density source of calories, improves the palatability of foods, and is needed to maintain normal essential lath, acid status. In general, CF patients with pancreatic insufficiency are not at risk for developing hyperlipidemia. (145) However, patients with pancreatic sufficiency may be at risk and should be screened according to national guidelines for the general population. (146)

Individuals with CF should be encouraged to follow a normal dietary pattern with no specific restrictions. The dietitian can assist patients in selecting more energy-dense foods with additional snacks to improve energy intake. Snacks such as nuts, muffins, cheese, and milkshakes may be helpful. Commercial oral supplements also may be used, but one must ensure that these costly supplements are an addition to, rather than a replacement for, calories from food.

Careful monitoring of patient nutritional status is aimed at the early detection and correction of unfavorable trends. Patients should be made aware of their ideal body weight (IBW) range, as estimated by using the Metropolitan Life Insurance Company height and weight tables for individuals of small frame. (147) Instructions can be provided for patients to monitor their weight at home and to report promptly significant weight loss. Weight should be measured on each clinic visit and compared to IBW based on height. The Clinical Practice Guidelines for Cystic Fibrosis (1) of the CFF suggests that patients be categorized as adequately nourished (> 90% IBW), underweight (85 to 89% IBW), mildly malnourished (80 to 84% IBW), moderately malnourished (75 to 79% IBW), or severely malnourished (< 7.5% IBW). Some centers track body mass index (BMI); BMI is calculated as follows: weight (kg)/height ($(m.sup.2)$) with a normal range of 20 to 25. A BMI of < 19 indicates significant malnutrition and a need for aggressive nutritional intervention. Some centers also find it helpful to measure triceps skinfold and mid-arm circumference as indicators of body fat and lean body mass. (133)

Approach to the Malnourished Patient: Patients who are malnourished or are losing weight should be evaluated in more detail and observed more closely. A dietitian should assist in the evaluation. Caloric need and actual intake should be assessed. Energy needs are based on individual requirements and can be calculated based on basal metabolic rate using the World Health Organization equations. (133) It is also important to assess malabsorptive symptoms. The approach to patients who continue to have steatorrhea despite taking appropriate doses of enzymes has been reviewed above. For patients with weight loss unrelated to caloric intake or malabsorption, alternative GI diagnoses and diabetes mellitus (DM) should be considered in the differential diagnosis.

Patients with moderate or severe malnutrition are candidates for more aggressive nutritional interventions. There are data suggesting that nutritional repletion has a positive impact on the course of the disease for such patients. (148-151) The placement of a nasogastric tube each night for supplemental feedings during sleep is an option for some patients, but this is inconvenient and poorly tolerated by some patients, particularly those with severe pulmonary disease. The insertion of a gastrostomy or jejunostomy tube is an acceptable method for increasing caloric intake in selected patients. Placement is generally well tolerated; however, the risk/benefit ratio must be carefully considered in patients with severe pulmonary disease because of the potential for respiratory compromise following the procedure. Choice of an enteral supplement must be individualized. Semi-elemental formulas do not require pancreatic enzymes, but complete formulas are generally well tolerated when given with enzyme therapy. (152) Parenteral nutrition may be appropriate for short-term nutritional repletion in a severely malnourished patient, (153) but the

enteral route is more appropriate and safer for long-term support.

Vitamin Supplementation: Patients with pancreatic insufficiency are prone to malabsorption of the fat-soluble vitamins (ie, A, D, E, and K). Clinicians must be aware that patients may manifest clinically important vitamin deficiency states (eg, night blindness in vitamin A deficiency, spinocerebellar degeneration or hemolytic anemia in vitamin E deficiency, metabolic bone disease in vitamin D deficiency, and bleeding diathesis in vitamin K deficiency). Vitamin supplementation is recommended, including the following: vitamin A, 10,000 IU/d; vitamin E, 200 to 400 IU/d vitamin D, 400 to 800 IU/d and adequate sunlight exposure; and vitamin K, 2.5 to 5 mg/wk. (133) The vitamins containing vitamins A, D, E, and K that are specially formulated for CF patients are sufficient for most adult patients when taken at a dosage of two tablets per day. Some patients require additional supplementation. To ensure adequate amounts of vitamin K, patients receiving frequent courses of antibiotics or those with a history of hemoptysis should be given an additional 2.5 to 5 mg vitamin K each week. (154) Serum levels of retinol, vitamin E, and 25-hydroxyvitamin D (25-OHD) should be checked annually, and vitamin doses should be adjusted accordingly. Clinicians should be aware that other nutrient deficiency states (eg, zinc, essential fatty acids, and antioxidants) have been reported in CF patients, but routine monitoring or supplementation is not recommended at this time. As in the general population, menstruating women often need supplemental iron to prevent iron-deficiency anemia. Other patients with CF are also at risk of developing iron-deficiency anemia. In some cases, iron-deficiency anemia must be distinguished from anemia resulting from chronic disease.

CF-Related DM

A CFF consensus conference (155) addressed the diagnosis, screening, and management of CF-related DM (CFRD). This section will briefly highlight the salient points of that report. Glucose intolerance and CFRD are age-related complications of CF. The CFF patient registry of > 22,000 individuals indicates that the incidence of insulin-requiring DM in CF-affected children < 10 years of age is similar to that of unaffected children (ie, < 1%). However, from adolescence into adulthood there is a progressive increase in the incidence of CFRD. More than 15% of patients > 35 years of age have CFRD and are receiving insulin therapy. (2,156) An even larger percentage, may have undiagnosed CFRD. Using oral glucose tolerance tests (OGTT) to screen a large patient population, one US CF center has reported (155,156) a prevalence of 43% in patients > 30 years of age (Fig 2). The pathogenesis of CFRD is complex, but largely is related to fibrosis and the destruction of the pancreas. Therefore, CFRD is seen most commonly in individuals who have exocrine pancreatic insufficiency.

(FIGURE 2 OMITTED)

CFBD is associated with increased morbidity and Mortality.

(144,157,158) Several studies (155) have shown deterioration in pulmonary function and BMI in the 2 to 4 years preceding the diagnosis of CFRD. In an analysis of 3,227 adults who were observed in the CFF patient registry from 1991 to 1996, the presence of CFRD requiring insulin was associated with more than a 2.5-fold increase in the relative risk of mortality during the observation period. A cause-and-effect relationship between CFIRD and excess morbidity and mortality cannot be definitively established. CFRD simply may be a marker of advanced disease and concomitant metabolic stress. However, one study (159) showed that the initiation of insulin therapy reversed the deterioration in body mass and pulmonary function in patients with CFBD. This lends weight to the hypothesis that untreated CFRD is an independent risk factor for poor outcomes. Also in favor of this hypothesis are the recent data showing that patients with relatively mild lung disease and impaired glucose tolerance appear to be at risk for a more rapid decline in pulmonary function compared to patients with normal glucose tolerance. (160) Microvascular complications occur in patients with

CFRD, with a reported incidence of 5 to 16% for retinopathy, 3 to 16% for nephropathy, and 5 to 21% for neuropathy. (161-163) However, the occurrence of macrovascular complications (eg, atherosclerosis, myocardial infarction, and stroke) appears to be rare. (164)

Several clinical situations that are more common in the adult patient appear to be associated with CFRD. Women with CF who become pregnant may be at increased risk of gestational diabetes. Of note, women with CFRD who become pregnant appear to be at increased risk of deterioration in pulmonary function and excess mortality in the 2 years following parturition compared to nonpregnant CF women with CFRD. (165) CFRD frequently develops in those patients with end-stage lung disease and has been associated with frequent pulmonary exacerbations, the use of corticosteroids, and supplemental (enteral or IV) feedings. The posttransplantation immunosuppressive regimen is frequently associated with onset and/or exacerbation of CFRD.

Screening and Diagnosis: A casual, or random glucose determination should be performed annually in all adults with CF. A value < 126 mg/dL (7 mmol/L) is considered to be normal. A value of (greater than or equal to) 126 mg/dL is considered to be abnormal and warrants measurement of fasting blood glucose (FBG) level. A casual glucose value of > 200 mg/dL (11.1 mmol/L) on two or more occasions is diagnostic of CFRD, as are either an FBG level of (greater than or equal to) 126 mg/dL on two or more occasions or an FBG level of (greater than or equal to) 126 mg/dL in association with a casual glucose level of > 200 mg/dL. (155) In ambiguous cases, an OGTT (using 1.75 g/kg up to a maximum of 75 g) should be performed. A 2-h postprandial glucose value of > 200 mg/dL is considered to be diagnostic of CFRD. An OGTT also should be performed in all women with CF who are contemplating pregnancy. It should be repeated soon after conception, and early in the second and third trimesters. (166) Certain clinical situations such as unexplained weight loss or failure to gain weight, delayed onset of puberty, and unexplained deterioration in pulmonary function suggest the possibility of undiagnosed CFRD. Glucose tolerance should be assessed in these situations.

Treatment of CFRD: The consensus of the committee was to treat CFRD aggressively in patients with fasting hyperglycemia. The principles of insulin and nutritional therapy in CFRD differ from those in either type 1 or type 2 DM. Therefore, the optimal management of CFRD is by the multidisciplinary CF team in conjunction with an endocrinologist or diabetologist. The goals of therapy are as follows:

- * Maintain optimal nutritional status;
- * Control hyperglycemia to reduce acute and chronic diabetes complications;
- * Avoid severe hypoglycemia;
- * Promote psychological, social, and emotional adaptation to living with CFRD; and
- * Be as flexible as possible within the framework of the patient's lifestyle.

Patients with CFRD and fasting hyperglycemia should be treated with insulin. The use of oral hypoglycemic agents cannot be recommended at this time. The nutritional management of CFRD is similar to the general approach for all patients with CF except that more attention is paid to the timing of meals and the avoidance of concentrated carbohydrates. The pattern of hyperglycemia in CFRD differs from that in type 1 DM. Some basal insulin secretion is usually preserved, making fasting hyperglycemia less severe and ketosis extremely uncommon. In contrast, postprandial hyperglycemia is often a prominent feature of CFRD. Therefore, a typical regimen often includes the use of very short-acting insulin (eg, lispro) prior to each meal. Appropriate dosing requires that patients either eat at predictable meal or use a carbohydrate counting system to estimate insulin requirements. A small amount of long acting insulin may be required as

well. Regular home glucose monitoring is essential in making the necessary adjustments in the insulin regimen.

Monitoring of therapy also should include the quarterly measurement of hemoglobin (A_{1c}). Hemoglobin (A_{1c}) values are often a less useful guide than in type 1 DM, but similar target values of < 7% for adults and < 8% for adolescents should be the goal. However, even a "good" glycohemoglobin value may be associated with an unacceptable degree of post-prandial hyperglycemia in some individuals. Goals for glycemic control in pregnant women are more stringent than those for men and nonpregnant women. (167)

All patients with CFRD should be screened annually for microvascular complications with a dilated eye evaluation and a urinalysis for microalbumin measurement. The presence of proteinuria should be taken particularly seriously in patients with CFRD, as it may indicate the onset of diabetic nephropathy. The concomitant use of nephrotoxic drugs (eg, aminoglycoside antibiotics, nonsteroidal anti-inflammatory drugs, and posttransplant immunosuppressive agents) may increase the susceptibility to nephropathy. As with all people with diabetes, hypertension should be aggressively treated, usually with a regimen that includes an angiotensin-converting enzyme inhibitor.

The natural history of the abnormalities in glucose metabolism in CF patients is not fully understood. At this point in time, the treatment for impaired glucose tolerance and CFRD without fasting hyperglycemia is not recommended except in the context of clinical trials or in the clinical situations detailed above (eg, unexplained weight loss). (155)

The diagnosis of diabetes is difficult for some adults who see it as yet another burdensome imposition or as a sign of end-stage disease. Monitoring and treatment of CFFRD are relatively labor-intensive and add to an already complex regimen. The support and understanding of the CF team will help the patients to incorporate this additional challenge into their daily regimen.

Hepatobiliary Disease

A CFF consensus conference (168) addressed the clinical features, diagnostic evaluation, and management of liver and biliary disease. The salient features of that document are highlighted here. The involvement of the liver and biliary tree in CF may lead to a gradually progressive biliary fibrosis and cirrhosis. It is estimated that up to 17% of children have clinically significant liver disease. (169,170) Data about the prevalence in adults are incomplete, but in one retrospective review (170) of 233 adults (> 15 years of age), 24% were found to have hepatomegaly or persistently abnormal liver blood test results. As the median survival time for CF patients increases, clinically significant liver disease and its complications will likely become a more important consideration.

Screening for Liver Disease: Examination and measurement of the liver and spleen by palpation and percussion should be performed at each clinic visit. A panel of liver function tests (LFTs) including serum aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, (gamma)-glutamyl transferase (GGT), and bilirubin should be obtained yearly. None of these tests correlates with the degree of hepatic fibrosis. Nevertheless, if any of these values is > 1.5 times the upper limit of normal, repeat testing should be performed within 3 to 6 months. Patients with persistent elevations (ie, occurring for > 6 months) or high elevations (ie, more than three to five times the upper limit of normal) of LFT results should be investigated more completely.

Diagnostic Evaluation: A workup for liver disease should begin with a focused history and physical examination. Other causes of elevated aspartate aminotransferase and alanine aminotransferase levels (eg, hepatitis A, B, or C virus; cytomegalovirus; Epstein-Barr virus; alcohol; drugs; or toxins) and/or elevated GGT or alkaline phosphatase levels (eg, gallstones, cholecystitis, biliary obstruction, or bone disease) should be

considered in the differential diagnosis. In addition to the routinely measured LFTs, biochemical evaluation should include total and direct bilirubin levels, total protein level, albumin level, prothrombin time, blood ammonia level (if significant portal hypertension is suspected clinically), cholesterol and glucose levels, and a CBC count to check for hematologic consequences of hypersplenism.

An ultrasound evaluation of the right upper quadrant of the abdomen should be obtained to detect the presence of gallstones, common bile duct stones, nodularity of the liver suggesting cirrhosis, findings suggesting steatosis, and bile duct or hepatic vein dilatation. This test provides very useful information but cannot detect or quantify the degree of fibrosis of the liver. Hepatic scintigraphy has limited clinical utility in the evaluation of suspected liver disease.

Endoscopic retrograde cholangiopancreatography (ERCP) can reveal strictures, dilatation, stones, and other abnormalities of the biliary tree. Although changes in intrahepatic bile ducts are relatively common in CF liver disease, (171) common bile duct stenosis is much less common (ie, < 10% of patients with advanced liver disease) than initially reported. (170) Because of the risks associated with this procedure, ERCP should be used only when there are clear-cut clinical indications. MRI cholangiography is a noninvasive technique that is used to visualize the biliary tree. It may be useful in examining the extrahepatic biliary tree, gallbladder, and major intrahepatic ducts. (172) This imaging modality should be considered prior to the use of ERCP.

Upper GI endoscopy is the most sensitive way to detect esophageal varices, gastric varices, portal hypertensive gastropathy, or gastric and duodenal ulcers. Endoscopy should be considered in adults with portal hypertension to determine whether varices are present since this information has prognostic and therapeutic implications.

Liver biopsy may be useful in making a specific diagnosis, and in determining the presence and extent of portal fibrosis or cirrhosis. However, not all clinicians believe that liver biopsy is indicated in investigating liver disease in CF because of the patchy nature of CF-related biliary cirrhosis and the lack of definitive therapy. Careful consideration of the risks of a liver biopsy should be made on an individual basis in conjunction with an experienced gastroenterologist.

Management of Liver Disease: A multidisciplinary approach is recommended for the management of liver disease in CF. The team should include the CF center staff, a gastroenterologist/hepatologist, a surgeon experienced in hepatobiliary surgery, and a radiologist. The reader is referred to the 1999 CFF consensus document (168) for a detailed description of each of the following three major CF-related causes of liver disease: (1) cholestasis/biliary cirrhosis/multilobular cirrhosis; (2) hepatic steatosis; and (3) hepatic congestion from cor pulmonale. It is important to establish a definitive diagnosis since each is managed differently and is associated with its own set of complications. For the purposes of this document, we will focus on the most common CF-related liver lesion, cirrhosis. Cholestasis, focal biliary cirrhosis, and multilobular cirrhosis are part of a sequential progression that occurs over a variable period of time, therefore, certain aspects of treatment are similar for these three lesions. The therapeutic approach includes medical therapy, nutritional therapy, management of complications (eg, portal hypertension and liver failure), and prophylactic therapy.

MEDICAL THERAPY: There is convincing evidence of the benefit from therapy with ursodeoxycholic acid (UDCA) in non-CF patients with a similar liver lesion (ie, primary biliary cirrhosis). (173,174) UDCA therapy significantly retards the progression of this cholestatic liver disease, as evidenced by improved survival times free of liver transplantation. UDCA therapy improves bile flow in CF patients, (175) may displace toxic hydrophobic bile acids that accumulate in the cholestatic liver, (176) may

have a cytoprotective effect, (176,177) and may stimulate bicarbonate secretion into bile. (178) Several, but not all, clinical trials (179-185) have demonstrated improvement in LFT results with UDCA therapy. A randomized, double-blinded, placebo-controlled trial (186) demonstrated significant clinical, nutritional, and biochemical improvement in the group treated with UDCA. Liver histology improved in another prospective 2-year trial of UDCA treatment. (187)

Conclusive evidence that UDCA alters mortality or the progression to cirrhosis is lacking. Nonetheless, clinicians should strongly consider treating CF patients who have cholestasis/fibrosis/cirrhosis. The appropriate dose is 20 mg/kg/d UDCA administered in two daily doses. (188,189) Adverse effects and toxicity from UDCA are uncommon. When they occur, they are usually not clinically significant and rarely lead to the discontinuation of treatment. Monitoring during therapy should include LFTs 3 months after initiating therapy and each 6 to 12 months thereafter. Repeat liver biopsy is generally not recommended because the focal nature of the liver lesion may make the assessment of histologic change over time difficult in an individual patient. There is no scientific justification for using UDCA in patients with CF who have little or no documented liver dysfunction or portal fibrosis.

Taurine has been suggested as an adjunctive therapy in liver disease because CF patients are commonly deficient in taurine. (185) However, in a randomized, double-blind trial, Colombo et al (186) showed no significant effect of taurine supplementation (17 to 33 mg/kg/d) on liver blood test results or fecal fat excretion measurements in patients with CF liver disease who were treated with UDCA or placebo.

NUTRITIONAL THERAPY: Monitoring fat-soluble vitamin status is even more important in the presence of liver disease (190) than in the presence of pancreatic insufficiency alone. All vitamin doses should be given with a meal and with pancreatic enzyme supplements. Supplementation with the water-soluble form of vitamin E (d-(alpha)-tocopherol polyethylene glycol (PEG)-1000 succinate) at a dose of 400 to 1200 IU/d will correct or prevent vitamin E deficiency in this setting. Patients also may need large doses of vitamin (D_{sub}2) or (D_{sub}3) (800 to 1,600 IU/d) or 25-OHD (calcifediol; 2 to 4 mg/kg/d) to normalize serum 25-OHD concentrations. The optimal dose of vitamin A supplementation has not been determined; however, patients with low serum retinol levels (ie, < 15 to 20 (micro)g/dL) should be supplemented with 10,000 to 20,000 IU per day. Serum retinol and retinol-binding protein should be monitored to ensure the adequacy of therapy, as well as serum retinol ester concentration to assess for toxicity (elevated serum retinyl esters). Prothrombin time should be monitored to assess vitamin K status. Significant prolongation of the prothrombin time should be treated with 5 to 10 mg vitamin K given once per week to daily, depending on the response to therapy. Following any change in vitamin dosing, repeat biochemical testing to ensure nutritional adequacy should be performed in 1 to 2 months.

PORTAL HYPERTENSION AND LIVER FAILURE: A detailed description of the management of these complications can be found in the CFF Consensus Statement. (168) If esophageal varices are documented by endoscopy, prophylactic (beta)-blocker therapy should be considered. There is no direct evidence of efficacy in CF, but (beta)-blockers reduce the risk of a first episode of variceal bleeding (191-193) in adults with other forms of liver disease. Adverse effects of (beta)-blockers, such as bronchospasm and depression, need to be taken into consideration. Endoscopic variceal ligation can be considered for patients who have large varices or who do not tolerate (beta)-blocker therapy. (194,195) Patients who are at high risk of bleeding from varices or have recurrent variceal bleeding may be candidates for a portosystemic shunt. (196,197) Liver transplantation is a viable consideration for a patient with decompensated cirrhosis, particularly if pulmonary function is relatively well-preserved. Liver

transplantation in CF patients results in a 1-year survival rate of approximately 75 to 80%. (198,199)

PROPHYLACTIC THERAPY: All patients with CF liver disease should receive a complete immunization series for both the hepatitis A and hepatitis B virus vaccines, unless prior infection with these viruses has been documented. Patients should be counseled about the risks of ethanol use and encouraged to avoid ethanol. Potentially, hepatotoxic medications and herbal therapies also should be avoided, if possible.

Other Hepatobiliary Manifestations: Micro-gallbladder occurs in about 30% of patients, and cholelithiasis or cholecystitis occurs in 1 to 12% of patients. (200) Cholelithiasis in CF is not responsive to UDCA therapy. (201) Hepatic scintigraphy may be helpful in evaluating a patient for suspected cholecystitis. If, in the presence of gallstones, clinical symptoms of gallbladder dysfunction or pain are present or if LFT results remain abnormal, a laparoscopic or surgical cholecystectomy should be performed, unless end-stage liver disease is present. The health-care team should be aware of the potential impact of any abdominal surgery on pulmonary function, particularly in patients with moderate-to-severe pulmonary dysfunction. A liver biopsy and intraoperative cholangiogram always should be obtained during any cholecystectomy procedure in a patient with CF.

Common bile duct stenosis occurs occasionally in patients, possibly because of compression of the bile duct by the fibrotic pancreas. (170,171) Primary sclerosing cholangitis may occasionally occur; however, cholangiographic findings consistent with primary sclerosing cholangitis may be caused by CF. Finally, cholangiocarcinoma also has been reported.

(202)

GI Complications

Abdominal Pain: Abdominal pain is a frequent complaint in patients with CF. (203) Of the many causes of abdominal pain, only distal intestinal obstruction syndrome (DIOS) and fibrosing colonopathy are unique to CF. All other causes of abdominal pain that occur in the general population also occur in patients with CF, and do so with the same presenting signs and symptoms. However, some causes of abdominal pain that are unusual in the general population occur at an appreciably higher frequency in CF. Hence, the list of likely diagnoses that need to be considered when evaluating a patient with CF and abdominal pain is longer than it otherwise would be.

A practical approach to abdominal pain in a patient with CF is to build a differential diagnosis based on the primary location of the pain (ie, epigastric, periumbilical, or hypogastric). The relatively frequent causes of epigastric pain in CF are gastroesophageal reflux, biliary tract disease, pancreatitis, and gastritis/peptic ulcer disease. Periumbilical pain can be caused by DIOS, appendicitis, and intussusception. Hypogastric pain in CF can arise from DIOS, C difficile colitis, and, much less commonly, from fibrosing colonopathy or colon cancer.

EPIGASTRIC PAIN: In a survey of 50 adults with CF, a majority had symptoms of gastroesophageal reflux. Eighty percent had history of heartburn, and 56% had a history of dyspepsia. (204) The evaluation and management of reflux in CF patients does not differ from that of the non-CF population, other than possibly avoiding postural drainage positions during CPT that exacerbate reflux. Cholelithiasis is another source of epigastric pain occurring in up to 12% of patients with CF. Alkaline phosphatase and GGT are often elevated in patients with CF because of intrinsic liver disease, with the result that these tests are less specific for cholelithiasis in this group. Diagnostic evaluation and treatment of cholelithiasis in CF can be found in a recent review by a CFF hepatobiliary disease consensus group. (168) Because of the relatively high frequency of biliary tract disease in CF, an abdominal ultrasound is often helpful in evaluating epigastric or right-upper-quadrant abdominal pain in CF patients.

Pancreatitis is a cause of epigastric pain that occurs almost exclusively in the subgroup of patients with CF who are pancreatic sufficient. (205) The recognition that mutations in CFTR are found in a large subgroup of patients with chronic pancreatitis who lack other evidence of CF has complicated the perception of what constitutes a CF diagnosis. Pancreatitis in CF usually presents with acute recurrent episodes, but chronic abdominal pain also is seen. The approach to diagnosing and treating pancreatitis in CF patients is similar to that for the general population. Although the incidences of gastritis and peptic ulcer disease have not been reported to be increased in CF patients over the general population, they are still common causes of epigastric pain in CF patients. Their evaluation and treatment are also similar for the CF and the non-CF populations.

PERUMBILICAL PAIN: Perumbilical pain in adults with CF is most often caused by DIOS, which is discussed elsewhere in this article (see "DIOS" section). Appendicitis, presenting with the classic symptoms of perumbilical pain that migrates to the right lower quadrant, occurs in patients with CF, although probably at no greater frequency than in the non-CF population. (206,207) The diagnosis of appendicitis often may be delayed in CF patients because its early symptoms are mistakenly attributed to DIOS. Older studies in the literature (208) reported that intussusception in the region of the iliocecal valve was relatively frequent in adults with CF. However, common experience suggests that clinically significant intussusception is rare.

HYPOGASTRIC PAIN: Hypogastric pain and abdominal bloating are frequent complaints in patients with CF. The most frequent cause is malabsorption that allows ingested nutrients to reach the colon where bacteria metabolize them to gaseous products. An indication that malabsorption is the cause of the abdominal pain is the coexisting symptom of steatorrhea. The therapeutic approach is to optimize pancreatic enzyme supplementation, as reviewed above.

The other major cause of hypogastric pain in patients with CF is DIOS. The likelihood that DIOS is the source of the abdominal pain is increased if the patient has intermittent constipation and abdominal distension (see "DIOS" section).

A less common cause of hypogastric pain in CF is C difficile-associated colitis. (208,209) The diagnosis should be considered particularly if the patient recently has received antibiotics or if the pain is accompanied by fever, leukocytosis, and/or blood in the stools. However, positive test results for C difficile also have been described in individuals with CF with abdominal pain who lack these other signs and symptoms. (210) Positive stool culture findings and/or toxin can sometimes be found in asymptomatic patients with CF, confusing the issue further. When the diagnosis of abdominal pain remains uncertain, colonoscopy showing pseudomembranes and/or inflamed mucosa, or an abdominal CT scan showing colonic thickening will support the diagnosis of C difficile colitis.

OTHER CAUSES: Other causes of abdominal pain in adults with CF deserve comment, although their incidence is quite low. (211) Fibrosing colonopathy, which is associated with the ingestion of very high doses of pancreatic enzymes, is manifested by inflammation and strictures in the right colon. Although it has been reported to occur in adults with CF, (212,213) it is a syndrome restricted mostly to the pediatric age group. An association between Crohn disease and CF has been suggested in the older literature. (214) However, some of these cases may have been caused by fibrosing colonopathy, which was just being described at that time. Colon cancer also has been reported (202) to occur at increased frequency in CF, although its incidence is still quite low. Consideration of the diagnosis is often delayed because its signs and symptoms are initially attributed to more frequent conditions that have similar clinical manifestations. (215) Colon cancer screening recommendations for adults with CF have not been

established.

DIOS: Recurrent episodes of intestinal obstruction occur in 3.5% of patients with CF, (2,216,217) The obstruction is caused by excessively viscid intestinal contents as a result of CFTR mutations leading to abnormal digestive fluid secretion. The previous name, meconium ileus equivalent, has been replaced by DIOS in recognition that the site of obstruction can occur within the right colon as well as in the terminal ileum. (218) DIOS occurs almost exclusively in patients with pancreatic insufficiency. A number of precipitating factors have been suggested to be responsible for triggering episodes of DIOS, including dehydration, use of medications that suppress intestinal motility such as narcotics, and noncompliance with pancreatic enzyme replacement. However, in the majority of patients presenting with DIOS, no precipitating factor can be identified. (219)

The diagnosis of DIOS is based heavily on eliciting the symptoms of distal small bowel/right colon obstruction. Depending on the degree of obstruction, typical symptoms are decreased stool output, colicky periumbilical and/or right-lower-quadrant pain, abdominal distension, nausea, and vomiting. A physical examination may uncover high-pitched bowel sounds with rushes that can progress to silence if peritoneal irritation and secondary ileus occur. Occasionally, a mass in the right lower quadrant can be palpated, representing a distended cecum and right colon. If blood tests show a WBC count that is elevated above baseline, this suggests that bowel viability is jeopardized. Alternative diagnoses such as appendicitis and C difficile colitis also should be considered in this circumstance. Supine and upright radiographs of the abdomen usually reveal a right colon distended with bubbly-appearing intestinal contents and dilated loops of small bowel containing air-fluid levels. Although DIOS is the most common cause of these signs and symptoms, other less frequent diagnoses must be considered (see "GI Complications" section).

The goal of DIOS management is to recognize the condition promptly and to institute treatment early so as to avoid the need for surgical intervention, if at all possible. One of the greatest dangers is if DIOS is mistakenly diagnosed as irritable bowel syndrome. Unlike this latter benign condition, DIOS can rapidly progress to life-threatening total bowel obstruction. Treatment includes the correction of systemic dehydration that occurs secondary to vomiting and/or decreased oral intake because of abdominal pain. If the patient is not vomiting and bowel sounds are present, a cautious attempt at relieving the obstruction by an oral or nasogastric tube route can be made. Although a variety of agents used to thin bowel contents has been recommended, such as diatrizoate (a radiographic contrast solution) (220) or N-acetylcysteine, (221) PEG electrolyte solutions are now frequently employed. (222,223) If the severity of bowel obstruction is too great to safely administer treatment by mouth, enemas can be used in an attempt to relieve the blockage. Large volumes of lavage fluid are sometimes required to clear the bowel. The success of clearing the right colon by enemas may be improved if radiocontrast dye is added to the solution and the procedure is monitored fluoroscopically. (224) At the first signs that the obstruction is lessening and that bowel motility, is returning, a PEG solution can be administered by the oral or nasogastric route. However, if signs and symptoms warn that the integrity of the bowel wall is in imminent danger, prompt laparotomy is needed to resect a questionably viable bowel and to relieve the intraluminal obstruction by irrigation.

A small number of patients with CF experience recurrent episodes of DIOS and require preventive therapy. Chronic oral administration of N-acetylcysteine, mineral oil, prokinetic agents, (225) and, more recently, PEG solutions have been recommended, if the episodes occur infrequently, the institution of aggressive treatment at the first sign of blockage may be sufficient. In the rare circumstance in which DIOS recurs frequently, a

daily PEG solution can be prescribed with the dose titrated to relieve symptoms.

Cancer Risks and Screening

Cancer risk becomes a concern as the population of individuals with CF ages. Although patients with CF appear to be at greater risk for some specific cancers, overall there appears to be little excess risk (if any) for cancer. The largest study conducted to date, (202) a retrospective cohort study of > 25,000 patients with CF in the United States and Canada, showed no increase or decrease in overall cancer risk. There was, however, a clear increase in the risk of cancers of the digestive tract, where 13 cancers were observed and only 2 had been expected (observed/expected ratio, 6.5; 95% confidence interval, 3.5 to 11.1). These cancers were distributed throughout the GI tract, with primary sites in the esophagus, stomach, small bowel, and large bowel, as well as the liver and pancreas. An analysis from the United Kingdom (226) involving 412 patients demonstrated a similar increased risk of pancreatic and intestinal cancer. The pathophysiology underlying the increased risk of GI cancers is unknown, but may be another manifestation of the CF disease process. Increased risks of cancer have been seen in other disorders affecting the GI tract, including Crohn disease and celiac disease. (227) A possible decreased risk of melanoma and breast cancer has been postulated for patients with CF and CF carriers, but research to date has not been supportive of this hypothesis. (228,229)

The application of primary and secondary preventive measures to the CF population should be a priority of physicians caring for these persons. Primary preventive measures, including smoking prevention/cessation and healthy diets that include antioxidants, such as vitamin E and selenium, are possibly more important for individuals with CF than for nonaffected persons. Secondary preventive measures that are designed for early cancer detection also should be applied within the CF population. It has been suggested that the early detection of colon cancer through fecal occult blood screening may reduce colon cancer mortality by up to 30%. (230,231) The sensitivity and specificity of occult blood screening in the CF population is unknown, and the test may be less specific in CF patients than in the general population. Nevertheless, endoscopy should be considered in an individual with CF and a positive occult blood test finding. Other recommendations, including annual mammography in women after age 40 years, breast and testicular self-examination, and prostate cancer screening should be followed by patients with CF, similar to their use in the general population.

ADOLESCENCE

Adolescence is a challenging period, both physically and emotionally, for children and families, even in the absence of chronic illness. The additional challenges of CF may affect normal adolescent development. Similarly, the challenges of adolescence may impact the health of the young person with CF. The pediatric CF team needs to be aware of these challenges, to assess their impact on individual development, and to plan effective interventions that will facilitate the movement of the adolescent with CF into adulthood and an adult-care setting. A summary of adolescent physical and emotional challenges and the potential complicating features of CF is listed in Table 3.

SITE OF CARE

Care for teenagers and adults with CF should be provided by personnel who are sensitive and responsive to their medical, developmental, and psychosocial needs. The model of a multidisciplinary team providing care for CF patients has been successful and should be incorporated into the care of adults with CF. Adults generally have more severe pulmonary disease, a higher prevalence of DM, and more complex financial and psychosocial issues. Therefore, a relatively higher intensity of pulmonary, respiratory, therapy, and endocrine, nutritional, and psychosocial services

may be needed. In addition, adult patients have unique needs, including vocational counseling, contraceptive and reproductive services, and obstetric care. There is consensus that a multidisciplinary team with training and experience in adult CF care should oversee the care of adults with CF. The multidisciplinary team typically consists of physicians with internal medicine training and additional expertise in CF, usually obtained as part of subspecialty training (eg, pulmonology), and nurses, dietitians, respiratory therapists, and social workers. Care is delivered in inpatient and outpatient facilities that are appropriate for adults, with laboratory services as outlined in the Clinical Practice Guidelines for Cystic Fibrosis. (1)

TRANSITION FROM PEDIATRIC TO ADULT CARE

Definition and Rationale

Obtaining health care in the adult care setting encourages independence and increased self-reliance. Transition should be a planned process over time, as an abrupt transfer to adult care could be unsuccessful. Important meetings and position statements of pediatric and adolescent health professionals in the past decade have brought consensus to the need for both adult care and a smooth transition for young adults with chronic conditions. (232-236)

Models of Transition

National and institutional policies, available financial and professional resources, and geographic proximity of CF pediatric and adult care clinics and hospitals all influence transition modeling for young adults with CF. The greatest distinction in choice of transition model is whether to transfer directly to adult care or to overlap pediatric and adult care over a period of time prior to transfer. The danger of the direct transfer approach is an abrupt severing of important relationships in pediatric care, with no introduction to the new care environment. This could lead to negative feelings for the patient and family, and ambivalence for the pediatric care team in letting go of their long-time patient. The likelihood of follow-through to the adult care setting could be seriously compromised. (237)

Young adults with CF have reported in satisfaction surveys that meeting with the pediatric physician and the adult care physician together prior to transfer to adult care is useful. (238-241) The joint meetings can be accomplished in either the pediatric or adult setting, in one or several clinic visits, depending on the patient's specific need, until the patient is seen alone in the adult care setting. (238,240,242-244)

Criteria for Successful Transition

There must be commitment by the institution trod by both pediatric and adult care teams to the importance of transition. There should be frequent formal and informal communication, and shared responsibility for developing coordinated and workable transition plans in order to ensure the continuity of the patient's care. (244)

Clinicians should introduce the concept of transition early, even as early as the time of diagnosis, when outlining the long-term care of CF. Children should be introduced to the concept of adult center care in an age-appropriate way. The adolescent can be given more responsibility for self-care and decision making, health education, and self-care training, and should be seen alone in clinic visits. (245) More intensive preparation can take place during the year before transfer to the adult clinic. This may include visits by adult care team members to the pediatric clinic, touring the adult care clinic, and discussions about doubts and anxieties. (246) Adult care physicians should be sensitive to the transition process and make an effort to know their new patient before transfer. Other strategies include inviting parents to the first visit in adult care (even though parents are not ordinarily involved in internal medicine clinics) and introducing any changes in medical treatment gradually, since abrupt changes can be perceived as disapproval of previous care. Transition

materials, such as adult program pamphlets, education packets, and readiness questionnaires are used in some CF centers to prepare patients and their families for adult care. (246-248)

Appropriate timing for transfer should be evaluated beginning in the high school years. In most cases, transfer will coincide with graduation from high school. The pace and progress of transition is expected to vary for individuals, depending on developmental maturity, self-care skills, special characteristics of the family, availability of adult clinicians and, in some cases, stage of illness. Those who are medically unstable, nearing death, or waiting for a lung transplant may defer transfer until stability is restored.

It is ideal to have a coordinator (eg, social worker or nurse) for the transition process. This person can ensure that a transition plan is created among the patient, family, and pediatric and adult care teams. The coordinator can schedule, facilitate, and track transition clinic and initial adult clinic appointments, and address any psychosocial issues of the patient, family, and clinicians throughout the transition process until the patient is adapted to adult care.

Evaluation

The ongoing evaluation of transition should occur at individual CF program sites to ensure the effective transfer of patients to the adult care setting. In general, there is a need for further study of transition models to identify which approaches work best in specific settings for specific patients and to evaluate whether the transition process helps individuals with CF to be successful in other areas of adult life.

EDUCATION AND CAREERS

Career Planning

Career planning is an important component of the ongoing care provided by the CF center. Ideally, at diagnosis parents should be informed that career planning will be necessary as their child grows older. The CF center team should initiate a discussion of careers with the adolescent patient and may need to assist the patient's school counselor regarding the implications of CF on career planning. The Meyers-Briggs type inventory, a career counseling tool, identifies jobs of interest that a person is capable of performing. Government-funded vocational rehabilitation programs are available in all states at no cost and provide job training, placement, and assistance with school tuition.

People with CF face few absolute restrictions on choice of employment. Education and career choices should be based on an individual's intellect, ability, interests, and life goals. The CF center team should tailor career counseling to the individual and should not make blanket exclusions of career choice. Present and future physical limitations should be considered.

Career Choice Considerations

In the United States, health insurance is primarily provided through employment. Employment decisions should take into account the benefits package offered by the employer. Employers who employ > 20 employees must offer health insurance as a benefit must provide Comprehensive Omnibus Budget Reconciliation Act (COBRA) continuation of coverage. Other employee benefits such as disability insurance coverage and life insurance should be reviewed. An employer who employs > 15 employees will be covered by the Americans With Disabilities Act (ADA).

Careers that allow for flexibility in work hours, reduced work hours, flexible use of vacation and sick leave, additional paid or unpaid sick time (including use of coworker-donated sick time), and the option of working from home are attractive. Self-employment often allows for maximum flexibility. Other considerations may include the levels of pulmonary irritants in a work environment, the particular stress of a job, and the potential exposure to upper respiratory viruses. Certain jobs, such as work with children, may pose a direct threat to the health of the individual.

with CF because of the increased risk of infection. Infection control issues may impact people with CF who choose careers in health care. The CF center team should educate such patients about the modes of transmission of infectious agents and the potential implications of employment in health care.

Postsecondary Education

Education itself can be a meaningful and life-enhancing experience. Those with the emotional, financial, and academic ability to handle college and postgraduate studies should be encouraged to pursue advanced education regardless of disease severity.

Determination of Financial Assistance Available: The CF center can assist the adult with information regarding the educational financial assistance that is available. Companies that manufacture and sell products to people with CF offer scholarships. In some cities, funds set up by families who have lost a loved one to CF also can be a source of financial assistance.

Selection of School: Students have a variety of choices when determining where and how to pursue a postsecondary education. Many new advances in technology have created the opportunity to take courses by teleconference or correspondence. Those interested in attending a school outside their home city should inquire about local CF care, state insurance assistance, and weather or environmental concerns.

Accommodations Needed by Student: Section 504 of the Rehabilitation Act of 1973 provides that no entity that receives federal funds can discriminate against a person on the basis of disability. Section 504 protects students at colleges from discrimination based on disability and requires accommodations needed by the student because of their disability. Many universities and colleges have an Office of Disabled Students that can assist the adult with CF. Typical accommodations for a student with CF may include the following: a private room; an air-conditioned room; a reduction in minimum hours required each semester; parking privileges on campus; and a plan for obtaining assignments or rescheduling exams because of absences. The CF center may need to provide documentation of the student's disability and the need for specific accommodations.

EMPLOYMENT

According to the 2000 National CF Registry, 2,684 adults with CF (31.0%) were working full time, 867 (10.0%) were working part-time; 1,675 (19.4%) were students, and 299 (3.5%) were homemakers.

Protection From Discrimination

The ADA, Title I, prohibits discrimination against a qualified individual with a disability in regard to all terms, conditions, and privileges of employment if the employer employs (greater than or equal to) 15 employees. The ADA also prohibits discrimination based on a relationship or association with an individual with a disability.

Accommodations for Disability

Under the ADA, people with CF also may be entitled to a reasonable accommodation if needed to allow them to perform the essential functions of their job. The provision of a reasonable accommodation can allow an adult with CF to continue working even if his or her health needs increase. Possible accommodations include additional sick time, modification of a work schedule, ability to work from home, and modification in policies and procedures of the employer.

Informing an Employer of a CF Diagnosis

It is illegal for a prospective employer to ask whether the applicant has a disability during the interview or on an application. Employers are allowed to inquire about qualifications, knowledge, skills, and ability to perform the job. After a person is hired, a person only needs to disclose a disability if the person needs a reasonable accommodation. The decision about disclosing a medical diagnosis can best be evaluated by the individual as he or she has knowledge of the specific work environment. The

CF center can provide information about CF to an employer. For some, the effort required to keep their health status a secret can be stressful, and disclosure may be best in such a situation.

Family Medical Leave Act

The Family Medical Leave Act provides that an eligible employee will be allowed 12 weeks unpaid leave per year if the employee has a serious health condition that makes the employee unable to perform the functions of his or her position. The time can be taken in a 12-week block or intermittently. A family member also may take leave to care for a child, parent, or spouse. The employee must work for an employer who has 50 or more employees, must have worked for 1 year for the employer, and must have worked 1,250 h in the prior year. The employer must continue health insurance coverage during the leave period.

MEDICAL INSURANCE

Obtaining and keeping medical insurance are two of the most important things for the adult with CF. In the United States, 85% of the population obtain health insurance through their employer, their spouse, or a parent's employer. If possible, changes in employment should be carefully planned to avoid interruption or loss of medical insurance.

The Health Insurance Portability Accountability Act

The Health Insurance Portability Accountability Act guarantees health insurance coverage in specific situations with relation to group health insurance for people who change or leave their jobs by providing creditable coverage that can be used to avoid a preexisting condition clause contained in the new policy as long as there is not more than a 63-day break in coverage. The Health Insurance Portability Accountability Act also mandates that if an employer provides health insurance to employees and/or dependents, then all employees and/or dependents must be covered regardless of their medical condition.

COBRA

COBRA requires that continuing medical insurance be made available to qualified individuals. The act applies to employers with (greater than or equal to) 20 employees who offer health insurance as a benefit. The maximum duration of continued insurance eligibility after employment termination depends on the circumstances (Table 4). In each situation, the employee must pay the premiums during the period of continuation of coverage.

State Law for Disabled Children

Most insurance policies contain limiting age language that ends insurance coverage for a dependent child when the child reaches a certain age. Most state insurance laws allow the child to continue as a covered dependent as long as the child demonstrates that he or she is incapable of self-support because of a physical or mental disability. The continuation of coverage will require that the treating physician sign a form indicating that the child meets the requirements of the law.

Denial of Coverage by an Insurer

It is becoming increasingly difficult for people with CF to receive approvals from their insurance companies for treatment needed. The adult with CF and the CF center team can be effective in advocating for the coverage of necessary treatment. Advocacy involves the appeal of a denial of coverage by the insurance company. Documentation of the medical necessity of the prescribed treatment will increase the likelihood of success of the appeal. If the appeal is denied, the CF center should not be discouraged but should appeal to the next level. In the event that Medicaid or Medicare denies treatment, the Medicare or Medicaid recipient also can appeal the denial of coverage.

DISABILITY INSURANCE

Benefits for disabled adults with CF may be available under the Social Security Disability Insurance (SSDI), Supplemental Security Income (SSI), Medicaid, and/or Medicare programs.

SSDI Benefits

SSDI benefits are provided to adults who have worked enough to qualify for benefits but are no longer able to engage in substantial gainful activity because of a physical or mental impairment. Qualification for SSDI does not involve the income level of the person applying for benefits, or their spouse or parent. SSDI provides a monthly benefits check after a 5-month waiting period, and Medicare benefits after 24 months of receipt of benefits.

SSI Benefits

SSI benefits are provided to adults who have not worked enough to qualify for SSDI, meet certain low-income guidelines, and are no longer able to engage in substantial gainful activity because of a physical or mental impairment. SSI is also available to children who meet the income and disability requirements. SSI provides a monthly payment aim Medicaid coverage effective immediately on approval of benefits.

Qualification for SSDI Benefits

Section 3.04 of the Social Security Listing provides that an adult with CF will qualify for SSDI benefits if the person has any of the following:

1. An FE(V.sub.1) equal to or less than the appropriate value specified ill Table 5, corresponding to the individual's height without shoes;
2. Episodes of bronchitis, pneumonia, hemoptysis (more than blood-streaked sputum), or respiratory failure requiring a physician's intervention, occurring at least once every 2 months or at least 6 times a year. Each inpatient hospitalization for > 24 h for treatment counts as an episode, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;
3. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased bacterial infection occurring at least once every, 6 months and requiring IV or nebulization antimicrobial therapy; and
4. If the person's condition is as severe as one of the listing requirements.

The CF center can be instrumental in the approval of an application for Social Security benefits. A letter from the treating physician indicating which section of the listing the applicant meets with attachments, such as PFT results or discharge summaries from hospitalizations, will significantly increase the chances that the adult with CF will be approved for benefits on the initial application. The reviewer may not have a medical background. If the applicant is denied benefits, an appeal must be filed within 60 days.

FAMILY PLANNING AND PREGNANCY

Fertility

Most men with CF are azoospermic because of anatomic abnormalities of the vas deferens and are functionally sterile, although 1 to 2% may be fertile. (249-251) Men in whom CF been diagnosed in adulthood who have mild mutations are more likely to be fertile. (252)

In contrast, the woman with CF has a normal reproductive anatomy. It is often stated that women with CF are less fertile than healthy women (253,254); however, > 100 women with CF become pregnant every year. (2) If fertility, in women with CF is decreased, it is unclear whether this is related to their general health status or is related to abnormalities of the cervical mucus. The rheology of the cervical mucus is different in the woman with CF compared to the nonaffected woman. There is lower water content and no thinning at ovulation. (253,255)

Contraception

Issues of female contraception are broadly similar for women with or without CF. (256) Men with CF should not assume that they are infertile. Semen analysis is recommended to determine fertility status. All persons with CF should exercise the same precautions as unaffected persons to

prevent the spread of sexually transmitted diseases.

Reproductive Decision Making

Genetic counseling should be offered to individuals with CF who are contemplating starting a family, and CF carrier screening should be offered to their partners. The couple should be advised of the potential effects of parenting on the health of the CF patient. Childbirth imposes long-term responsibility on parents, with or without the presence of CF. No one is guaranteed the opportunity to witness the growth of their children, but parents with CF must face the possibility of their own early death with more than just vague concern. It is imperative for health-care providers to recognize that they should not impose their own views on an individual patient's reproductive decision making, but rather that they should present the patients with medical information to allow them to make an informed decision. These issues should be addressed prior to conception. The woman with severe lung disease also should be made aware that panel-reactive antibodies may be increased during pregnancy. Some transplant centers may consider elevated levels of panel-reactive antibodies to be a contraindication to lung transplantation. (257)

Alternatives to Normal Conception

Alternatives to normal conception are available and should be considered on an individual basis. For the partners of infertile men, artificial insemination with donor sperm is an option. Microsurgical epididymal aspiration of spermatozoa with intracytoplasmic sperm injection into the oocyte may allow men with CF to become biological fathers, but it is available only in larger fertility centers and has a success rate per cycle in the order of 12 to 45%. (258-260) It is also an expensive process.

Pregnancy

Some reports demonstrate good outcomes for women with CF during and after pregnancy, (165) but there are also reports of untoward outcomes. (261, 262) There are normal changes that occur during pregnancy that may adversely affect the woman with CF. There are increases in minute ventilation (263) and oxygen uptake, (264) which may be problematic in the pregnant woman with severe lung disease. Blood volume and cardiac output can rise by as much as 50% toward the end of pregnancy as a result of the placental circulation and generalized vasodilatation. (264) These changes could precipitate right heart failure in the presence of severe lung disease. Thresholds of medical contraindication to pregnancy have not been established, although there are concerns about the patient with advanced disease, malnutrition, or diabetes.

Management of the Woman With CF During Pregnancy

Pregnancy in a woman with CF should be considered a high-risk pregnancy. It is important to provide continuity of comprehensive care by a coordinated team with knowledge and expertise in CF. In addition to the usual issues, such as monitoring of nutritional and pulmonary parameters, there are CF-specific issues that warrant careful attention, including altered drug pharmacokinetics. Screening for and treatment of DM during pregnancy is discussed below (see "CFRD" section).

An essential part of maternal care is the early recognition and prompt treatment of acute pulmonary exacerbations. If possible, medications with the least potential harm to the fetus should be chosen. There are several medications that are used for maintenance therapy, such as inhaled tobramycin and dornase-(alpha), which have a class C designation. The use of these medications should be determined on an individual basis.

Breast-Feeding

Breast milk appears to be normal in women with CF, including normal ionic concentration levels and normal levels of available nutrients. (265) Breast-feeding requires an additional intake of up to 500 kcal per day for the healthy mother (266) and appears to be well-tolerated by the woman with CF as long as she is able to meet the increased caloric demands.

BONE AND JOINT DISEASE

Bone Disease

Many individuals with CF experience bone and joint disease, including low bone mineral density (BMD). A CFF Consensus Conference on these topics was convened in 2002. Bone disease (ie, osteopenia or osteoporosis) may lead to kyphosis and fractures. These problems occur more commonly in adults and those who have undergone lung transplantation. (267-271) The prevalence of bone disease in patients with CF depends on the health status of the individual (including severity of lung disease and nutritional status) and the definition of bone disease. Low BMD has been widely reported in children and adults with CF, and is associated most closely with low BMI and low lung function. The prevalence of osteoporosis in CF adults varies from 38 to 77% (268,272-276) and is higher than that reported in children (19 to 67%). (272,277-280)

The pathogenesis of low BMD in CF patients involves both decreased levels of osteoblasts (ie, bone-forming cells) and increased levels of osteoclasts (ie, bone-resorbing cells). These data are supported by bone metabolism studies demonstrating accelerated bone resorption (ie, increased urinary cross-linked N-telopeptides of type-I collagen) and diminished bone formation (ie, low serum osteocalcin levels), (281) but further studies need to be conducted before a clear understanding of bone metabolism in CF evolves. Low vitamin D levels, found commonly in CF children and adults, (275,279,282,283) may contribute to reduced bone formation, but osteomalacia or diminished bone mineralization has not been well-documented. GI absorption of calcium is impaired in persons with CF as well. (284) Vitamin-D insufficiency may result from diminished sunlight exposure, poor vitamin D absorption, or accelerated vitamin D catabolism. Hypogonadism and low growth factor levels also may exacerbate low BMD. (280) Other, unidentified factors also may affect bone formation in CF. Accelerated bone breakdown probably results from corticosteroid exposure, diminished physical activity, and chronic pulmonary inflammation, with the latter mediated potentially by inflammatory cytokines. (285) After lung transplantation, an accelerated decline in BMD may result from immunosuppressant therapies or chronic posttransplant problems (eg, obliterative bronchiolitis).

Screening and Diagnosis: Dual-energy radiograph absorptiometry (DEXA) is a safe, accurate, fast, and inexpensive method of measuring BMD at the spine, femur, and other sites. Lateral chest radiographs can detect kyphosis, vertebral compression fractures, and osteopenia, but are insensitive to low BMD when compared to DEXA. Nonetheless, since chest radiographs are available on all patients, the presence of osteopenia, kyphosis, or vertebral fractures should encourage a more quantitative measure of bone mass by DEXA. As a general rule, children and adults with CF should be screened for bone disease by DEXA and, if BMD is normal (Z score (+ or -) SD) in children, 0 (+ or -) 1; and T score in adults, 0 (+ or -) 1), rescreening every 9. to 5 years may help to determine the rate of bone growth or loss. Z score, the number of SDs above or below age-matched control subjects, is used most often to define bone disease in children. T score, the number of SDs that a BMD measurement is above or below peak bone mass (which occurs between 25 and 30 years of age), is used most often to define bone disease in adults. For each SD below peak bone mass, the risk of fracture doubles. (286) Osteopenia is defined as a T score between - 1.0 and - 2.5, and osteoporosis as a T score < - 2.5. (287)

Treatment Recommendations: The treatment of established bone disease in CF patients has not been well-studied. Encouraging weight-bearing exercise, exposure to sunlight to promote vitamin D formation, maintaining a good nutritional status, and the proactive management of pulmonary infection are reasonable measures to maintain or increase BMD. Calcium and vitamin D supplementation is probably a useful intervention based on clinical studies in non-CF patients with low BMD. Calcium supplements in the form of calcium carbonate should be at least 1 g/d, an intervention

that does not increase (and may decrease) the risk of nephrolithiasis. Predicting the optimum vitamin D supplement is not easy because of the variability in absorption of vitamin D. (288) Vitamin supplements that contain vitamins A, D, E, and K probably are not sufficient when used alone if vitamin D deficiency exists in adults. (282) For vitamin D deficiency (ie, serum "25-OHD level, < 18 to 20 ng/mL), vitamin D supplementation should be individualized to obtain a serum 25-OHD level of > 18 to 20 ng/mL and preferably > 30 ng/mL. If osteopenia is present in adults, as defined above (or in children as evidenced by a Z score between -1 and -2.5), one should supplement with vitamin D to keep 25-OHD levels at > 18 to 20 ng/mL. Advice from an endocrinology consultation may be helpful to exclude sex hormone deficiency and other predisposing conditions. If Z scores in children are < - 2.5 or T scores in adults are < - .2.5 (the definition of osteoporosis) and/or minor trauma fractures or kyphosis occur, a more aggressive stance toward therapy should be initiated. Preliminary data suggest that pamidronate, an IV bisphosphonate, is very useful to remineralize bone in patients with CF who have had osteopenia, osteoporosis, or both before, (289) and after lung transplantation. (290) Bone pain and fever may occur in the former group, but not the latter group, limiting the usefulness of this therapy. In adults, therapy with oral bisphosphonates (eg, alendronate) or calcitonin may be useful based on anecdotal reports in CF and well-controlled trials in non-CF patients. The evaluation and treatment of patients with CF who are lung transplant candidates is of particular importance given the anticipated exposure to immunosuppressants. Fractures should be managed conventionally with immobilization and short-term opioid analgesics. Calcitonin may be particularly helpful for vertebral fracture-associated pain. Currently, multiple trials are underway to study different therapies for CF bone disease.

Joint Disease

Joint disease affecting adults with CF may present as episodic arthritis and/or hypertrophic pulmonary osteoarthropathy (HPOA). Arthropathy occurs in up to 12% of patients with CF, is more common in adults, and appears to be caused by immunologic processes. (291) Acute episodes may affect all joints, are usually asymmetric, present with swollen, hot, red, and tender joints, often cause incapacitating pain, typically last 7 to 10 days, and usually are not erosive. (292) Serologic analysis to exclude other causes of arthritis should be considered. Joint fluid analysis is usefully nonspecific and may be noninflammatory, but synovial tissue is often hyperemic and inflamed. (293-294) Short courses of nonsteroidal and steroidal anti-inflammatory medications are very useful in the management of CF arthritis. HPOA, which is similar to arthritis, is more common in adulthood (median age of onset 20 years), affecting approximately 8% of patients, and is characterized by chronic, proliferative long-bone periostitis, causing symmetrical bone pain and painful oligosynovitis in the large joints. (295-296) Unlike arthritis, HPOA exacerbations tend to accompany pulmonary infectious exacerbations. The etiology of HPOA is unknown. Radiographic findings, which are specific but not highly sensitive, include periosteal new bone formation at the distal ends of long bones. (297) Therapy with nonsteroidal anti-inflammatory agents is usually successful, but some patients require more potent analgesia.

END-OF-LIFE OPTIONS

Despite substantial therapeutic advances, CF remains uniformly fatal, and little has been written about end-of-life care for patients and their families. (298-299) Barriers to optimum end-of-life care for these patients include the following:

1. Difficulty predicting the timing of death. The most widely quoted predictive model (300) has been called into question. (144,301,302)
2. The influence of lung transplantation on decisions about

end-of-life care.

3. Personal fears or lack of confidence have prompted many physicians to avoid dealing with the reality of dying with its requirements for communicating "bad news" and prognosis, negotiating clear goals of care, and advance care planning. (303)

4. Fears of opiate addiction and exaggerated risks of adverse effects result in undertreatment/ inadequate control of symptoms in dying patients. (298,303)

Palliative Care

The amelioration of human suffering is an important goal for health-care providers. The attitudes, skills, and behaviors that physicians use to relieve suffering and improve quality of life for patients with life-threatening illnesses are termed palliative care. It may be combined with therapies aimed at restoring or prolonging life (eg, restorative care), or it may be the total focus of care. (303) An appropriate combination of restorative and palliative care is the hallmark of optimum management for patients with CF and their families. Palliative care includes, but is not limited to the following:

1. Interdisciplinary approach (involving the physician, nursing, social work, psychology, respiratory and physical therapy, pastoral care, and others).

2. Skilled management of symptoms that may cause suffering (eg, pain and dyspnea). This is the sine qua non of palliative care for patients with CF. Headaches and chest pain sharply increase in the 6 to 12 months prior to death in adults. (304) Opiates can be effective for pain and dyspnea without causing respiratory depression. (304-306) Issues such as anxiety, depression, and fatigue also require attention.

3. Efforts to maximize quality of life as defined by patient and family.

4. Family and care provider education, and training to improve patient care.

5. Assessment and treatment of psychological, social, and spiritual distress.

6. Respite care for family and care providers.

7. Assistance to plan for the last days of life, preparation of wills and other important documents, and planning for death, funeral, memorial service, burial, or cremation.

8. Loss, grief, and bereavement support for patient, family, and care providers.

Advance Care Planning

Planning is essential and should occur early in the course of the illness. The care team should be open, honest, and sensitive to family and cultural issues. Advance care planning is a process of structured discussion and documentation woven into the regular process of care. The goal is to ensure that a patient's wishes will be respected. It is a process that helps patients identify and clarify their personal values and goals about health, chronic illness, and medical treatment. The patient identifies the care they would like to receive in various situations. They also determine the person or persons whom they would like to make health-care decisions on their behalf in the event they cannot make decisions for themselves. (303) The sense of control and peace of mind that this process fosters in the patient and the family, and the anxiety reduction for proxy decision-makers are important benefits. (303) Most adults with CF have considered their wishes, often are aware of decisions made by others with CF, and are likely to be relieved when the physician discusses the issues with honesty, and sensitivity. Finally, the physician and CF team should be flexible and should offer choices for the location of terminal care and dying (ie, critical care unit, private hospital room, hospice, or home with requisite support).

Assisted Ventilation

Assisted ventilation for respiratory failure was thought of as futile for patients with CF at one time. (307) However, that has changed dramatically with the advent of noninvasive modes of ventilatory support (308-310) and lung transplantation. One report (311) has suggested that even patients with respiratory failure may benefit from ventilatory support and that this form of therapy should be considered, particularly in patients awaiting lung transplantation. The issue of whether and when to use mechanical ventilation in CF patients may complicate end-of-life care. End-of-life care may be more difficult in a critical care setting. While offering some hope, the care team also must be realistic in presenting options to patients and their families. At most centers, only a minority of patients will survive an episode of respiratory failure long enough to have the opportunity for lung transplantation. The withdrawal of ventilatory support may become a common terminal event for patients with CF.

Lung Transplantation

Lung transplantation became a viable option for CF patients > 10 years ago, but there is substantial morbidity and mortality associated with the procedure. (312) The 5-year posttransplant survival rate is approximately 50%. In addition, the cadaveric organ donor supply has lagged behind the number of potential transplant recipients, resulting in longer and longer waiting times. The inadequate supply of donor lungs has led to the development of the living donor, bilateral lobar transplant procedure (313) as an alternative to the standard cadaveric procedure. This procedure raises unique questions because of the potential morbidity and mortality to the healthy donors. However, if suitable donors are available, the major advantage of this approach is that it can be performed on patients who are unlikely to survive long enough on the waiting list to receive cadaveric organs. The limited data available suggest that posttransplant survival following this procedure may be shorter than that following the standard cadaveric procedure. (314)

The adult care team regularly faces difficult decisions about candidacy for transplantation. The rationale underlying selection criteria for lung transplant candidates has remained constant since the procedure was first performed. Patients are considered who have limited survival and have exhausted conventional therapies. The hope is that the procedure will prolong survival and improve quality of life. The International Society of Heart and Lung Transplantation and the American College of Chest Physicians have published guidelines with specific selection criteria. (315,316)

Since 1992, the selection of CF patients for transplantation has been heavily influenced by a survival model based on percent predicted FE(V.sub.1). The model suggested that 2-year mortality rate approaches 50% for CF patients with an FE(V.sub.1) < 30% of predicted. (300) It was recommended that clinicians consider referral of such patients for transplant evaluation. The survival implications of an FE(V.sub.1) of < 30% of predicted have been revisited more recently. (143,301,302) It is clear that FE(V.sub.1) alone is inadequate in identifying appropriate candidates for transplantation. A more precise, validated survival model (144,317) that incorporates several additional parameters can identify a subset of CF patients who will more likely gain a survival advantage from the procedure. These parameters (eg, gender, nutritional status, diabetic status, sputum microbiology, and number of pulmonary exacerbations) are readily available in the clinical setting. The CF care team should consider including this tool in their assessment of potential lung transplant candidates. Close communication between the CF care team and the transplant center is very important in identifying appropriate candidates for transplant, and in the choice and timing of the procedure.

Appendix: Participants

This list reflects current positions held by participants as of October 2002.

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Table 1--Complication Rates in Adults and Children *

Complication	Adult, %	Children, %
P aeruginosa colonization	79	47
Ciprofloxacin resistance	22	4
Tobramycin resistance	15	6
B cepacia colonization	6	2
Massive hemoptysis ((dagger))	1.8	0.1
Pneumothorax ((double dagger))	1.4	0.2
On supplemental oxygen continuously	6	1
CFRD requiring insulin	16.2	2.5

* 2000 CFF Registry-data.

((dagger)) Bleeding volume of > 240 mL in 24 h or requiring transfusion.

((double dagger)) Requiring chest tube.

Table 2--Phenotypic Features Consistent with a Diagnosis of Cystic Fibrosis

Chronic sinopulmonary disease manifested by:

Persistent colonization/infection with typical CF pathogens including *Staphylococcus aureus*, nontypeable *Haemophilus influenzae*, mucoid and nonmucoid *P aeruginosa*, and *B cepacia*

Chronic cough and sputum production

Persistent chest radiograph abnormalities (eg, bronchiectasis, atelectasis, infiltrates, and hyperinflation)

Airway obstruction manifested by wheezing and air trapping

Nasal polyps, and radiograph or CT scan abnormalities of the paranasal sinuses

Digital clubbing

GI and nutritional abnormalities, including:

Intestinal: DIOS and rectal prolapse

Pancreatic: pancreatic insufficiency and recurrent pancreatitis

Hepatic: chronic hepatic disease manifested by clinical or histologic evidence of focal biliary cirrhosis or multilobular cirrhosis

Nutritional: failure to thrive (protein calorie malnutrition), hypoproteinemia and edema, and complications secondary to fat-soluble vitamin deficiency

Salt loss syndromes: acute salt depletion and chronic metabolic alkalosis

Male urogenital abnormalities resulting in obstructive azoospermia

Table 3--Adolescent Challenges and Their Potential Interactions with CF

Challenges of Adolescence	Potential Impact of/on CF
Rapid physical growth	Delayed growth (height and weight gain) Decreased body fat (especially women) Prevalence of eating disorders may exceed that of the general population
Pubertal changes	Delay in genital development (male) Delay in menarche (female)
Sexual development	Delay in dating and sexual relationships Sexual dysfunction Misunderstanding of fertility Issues with reproduction Poor body image Low self-esteem and self-concept Parental overprotection Dependency
Development of personal identity	Difficulty forming intimate relationships Social isolation
Autonomy and independence	Fear of rejection leading to secrecy about illness Increased uncertainty of future Decreased expectations for self
Development of interpersonal relationships	Delay in planning for future Need for realistic planning Use of denial as coping strategy Sexual and substance abuse may be less than in general population, but still present Poor adherence to medications and therapy
Planning for the future	Delay in planning for future Need for realistic planning Use of denial as coping strategy Sexual and substance abuse may be less than in general population, but still present Poor adherence to medications and therapy
Risk-taking behavior	Use of denial as coping strategy Sexual and substance abuse may be less than in general population, but still present Poor adherence to medications and therapy

Table 4--Duration of COBRA Insurance Eligibility
Based on the Circumstances

Qualifying Event	Length of Period of Continuation, mo
Termination from employment	18
Death of the covered employee	36
Divorce or separation from covered employee	36
Covered individual has left work and become eligible for SSDI benefits	29
Individual has reached a limiting age on his or her parents' policy	36

Table 5--FE(V.sub.1) Criteria for SSDI Eligibility

Heights Without Shoes

cm	Inches	FE(V.sub.1), L
(less than or equal to) 154	(less than or equal to) 60	1.45
155-159	61-62	1.55
160-164	63-64	1.65
165-169	65-66	1.75
170-174	67-68	1.85
175-179	69-70	1.95
(greater than or equal to) 180	(greater than or equal to) 71	2.05

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Quality indicators for academic nursing primary care centers.
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Executive Summary
* Academic Nursing Centers (ANCs) provide a unique environment that serves patients, students, and faculty.
* As a result, performance measures of quality and efficacy must be broadly defined to meet their clinical, fiscal, research, and educational objectives.
* The University of Texas Health Services ANC defines measures within the following areas in their plan for performance management:
-- Multidisciplinary quality assurance committee and process
-- Financial stability
-- Billing and insurance systems
-- Participation levels from students and faculty in the ANC
-- Research activities
-- Patient care processes
-- Administrative organization
-- Marketing efforts
-- Medical information management
-- Credentialing and continuing education
-- Facilities and care environment
-- Health education and wellness services
-- Patient/corporate satisfaction feedback
-- Faculty and staff management

ACADEMIC NURSING centers (ANCs) involved in primary care provide a forum by which schools of nursing achieve a number of goals including community service, clinical opportunities for students and faculty, and data for research projects (Aydellote & Gregory, 1989). Regardless of the ANC's objectives, the issue of quality is paramount. The purpose of this article is to propose an evaluation tool for measuring the overall quality of a primary care ANC. Further, it is to stimulate thoughts on how to improve the overall quality of a primary care ANC clinic. It is not the intent to discuss quality care delivered by individual practitioners or quality related to specific health care conditions.

History of Academic Nursing Centers

The American Nurses Association's definition of a nursing center is a center in which a nurse occupies the chief management position; accountability and responsibility for client care and professional practice remains with the nursing staff; and nurses are the primary providers seen by clients visiting the center (Lockhart, 1993). Holthaus (1993) traces the beginnings of nursing centers back to Lillian Wald in 1893. Other examples of nursing centers through time have been the Frontier Nursing Service and community nursing centers funded by local, state, and federal monies, and by foundations such as Robert Wood Johnson and W.K. Kellogg. Planned Parenthood clinics most certainly fit the description of nursing centers, as do many college health services across the country. In the late 1970s Pace University in New York became one of the early leaders of ANCs.

Current accurate data on the number, diversity of missions, economic status, quality, and other issues of ANCs are not available. The National League for Nursing provided an excellent forum for those involved in ANCs in the late 1980s and early 1990s. However, that forum is no longer active. Important current forums for ANCs include the American Association of Colleges of Nursing (AACN) and the recently developed Penn Macy Fellowship program at the University of Pennsylvania. Nurse researchers involved with these two organizations are actively looking at ways to gather data that will identify the varied types and activities of ANCs.

Review of Literature on Quality of Care at ANCs

The literature is anemic and confusing with regard to quality issues in ANCs. However, Davis (1993) discusses the various issues of accrediting bodies for primary care settings (protocols, quality assurance, and risk management programs) and indicates that accreditation is one means to assure excellence of care in an ANC. Chickering Group (Ferran, 2001) raises the issue that the quality of a health care service is viewed differently by patients, providers, administrators, insurance/managed care companies, and in some cases students (student health services). Patients often define quality in terms of convenience and service expectations. Providers define quality in terms of process and outcomes. Administrators define quality as "no complaints" and cost effectiveness. Researchers typically evaluate quality of care by assessing the structures, processes, and outcomes of care (Rosenfeld & Wenger, 2000).

Definition of Terms

Just as patients, providers, and researchers measure and define quality in various ways, they also differ in the meaning of other terminology surrounding the issue of quality. For the purposes of this article the following terms are defined.

Quality refers to the degree of excellence. In this article it refers to the degree of excellence of a particular center, program, or other service pertaining to an ANC.

A quality indicator is a policy, program, protocol, standard, guideline, assessment measure, or other evaluation tool that shows there is reason to believe measures are in place to assure a high level of care is provided.

Quality assurance is defined by Mosby (Anderson, 1994) as "any evaluation of services provided and the results achieved as compared with accepted standards" (p. 1319).

A standard is an "evaluation that serves as a basis for comparison for evaluating similar phenomena or substances such as a standard for a practice" by a professional (Anderson, 1994, p. 1475). In this case we refer to standards not just for a professionals but also for ANCs. Standards and guidelines are developed over time by organizations and professionals who present the highest levels of achievement in a particular area such as clinical practice or health care management. Standards are also recommendations by which an individual or organization can achieve goals.

A quality assurance program is a systematic "review of selected

hospital medical/nursing records by medical/nursing staff members, performed for the purposes of evaluating the quality and effectiveness of medical/nursing care in relation to accepted standards" (Anderson, p. 1319).

Quality assessment is a method that can be used to obtain information related to the provision of health services that should subsequently lead to appropriate action to safeguard and enhance the quality of these services. Quality assessment is achieved by identifying and describing the structural characteristics of a setting, examining the activities or processes involved in the provision of services, and measuring the effects of services that result when the identified structures and processes are combined (Salazar, Graham, & Lantz, 1999).

A quality assessment measurement is a "formal, systematic, organizational evaluation of overall patterns or programs of care, including clinical, consumer and systems evaluation" (Anderson, p. 1319). Typically, quality measurements evaluate structures, processes, and outcomes.

National Organizations and Quality

Accreditation from a national organization such as The Accreditation Association for Ambulatory Health Care (AAAHC), the Joint Commission on the Accreditation of Healthcare Organizations (JCAHO), and the Community Health Accreditation Program (CHAP) may or may not be appropriate depending on the individual ANC's make up. Protocols, however, are frequently mandated by state law and, if properly written, will address health care issues from a "clinical standards" viewpoint. One example of a quality assurance program is that of Pace University Health Care Unit. This program includes peer review of charts, diagnosis-based chart review, review of high-risk client encounters, monitoring of equipment, systematic disposal of expiring medications, adherence to the Occupational Safety and Health Administration (OSHA) and Department of Environmental Control standards for environment, client satisfaction surveys, followup file for client surveys, and followup of missed appointments (Davis, 1993).

Four organizations that provide a framework for assessing quality in the primary care setting are AAAHC, JCAHO, CHAP, and the American Academy of Ambulatory Care Nursing (AAACN). AAACN first developed and published standards for ambulatory care nursing practice and administration in 1985 and has since revised them (AAACN, 2000). These standards come closest to defining appropriate quality indicators for ANCs. AAACN's nine standards are accompanied with rationale and measurement criteria by which any ANC could evaluate itself for quality issues. These standards are:

1. Structure and organization of ambulatory care nursing standards of professional performance.
2. Staffing standards of professional performance.
3. Competency standards of professional practice.
4. Ambulatory nursing practice standards of professional performance.
5. Continuity of care standards of professional practice.
6. Ethics and patient rights standards of professional performance.
7. Environment standards of professional performance.
8. Research standards of professional performance.
9. Quality management standards of professional performance.

Despite the comprehensiveness of these standards, they still do not fully address some of the more important quality indicators of an ANC.

Quality Indicator Recommendations

AAAHC, JCAHO, CHAP, and AAACN are all concerned with multiple areas of service (see Table 1). Although they address the typical quality indicators of primary care centers, none of them directly addresses the unique issues of academic nursing centers.

The following 14 recommendations are based on AAAHC, JCAHO, CHAP, and AAACN recommendations and guidelines with the addition of other quality indicators unique to academic practices. These recommended indicators are

based on 12 years of practice at The University of Texas Health Services ANC.

1. A highly qualified primary care academic nursing center will possess:
1. Quality assurance (QA) program. The QA program will have committee members representative of clinic personnel from various areas including administration, providers, and clerical staff. The committee will also be multidisciplinary. The program will be organized as evidenced by holding regular meetings, developing and implementing a plan of action, and systematically reviewing areas represented by its members. Finally, it will have and audit a compliance program addressing state and federal regulations (for example, Health Insurance Portability & Accountability Act or HIPAA, Medicare, Medicaid, and managed care organizations).

2. Financial stability. Financial stability will be evidenced by a collection rate above 90%; accounts receivable (AR) for greater than 120 days at less than 18% of total AR; ability to track accuracy of billing; financial self-sufficiency (income exceeds expenses); reserve funds equal to 10% of annual income; administrative costs no greater than the national mean according to the practice type (see national data from Medical Group Management Association); and diverse funding sources such that less than 50% of funding comes from a single source.

3. Educational opportunities for students. A specified number (or percent) of students receive educational opportunities in the center. Faculty from the school of nursing have the opportunity to use the ANC for faculty practice opportunities. Faculty preceptors receive orientation (Fay et al., 2001). At least 50% of clinic faculty/staff are significantly involved in teaching students. Clinic personnel are from different disciplines (for example, nursing, medicine, pharmacy, social work).

4. Research efforts. Faculty from the ANC and the academic unit are actively engaged in research and publishing. Students regularly use ANC clinical data for dissertations, theses, and clinical projects. Institutional review board protocols are followed. ANC resources (funds, space, computers, release time) are available for research purposes. ANC administration encourages interdisciplinary research efforts.

5. Patient care processes. The ANC has appropriate referral and consultation patterns (Mackey, Cole, & Veeser, 1999). Laboratory, radiology, physical therapy, and other outpatient care services are offered and monitored for quality and service. Clinic pharmaceuticals are regularly monitored for dates of expiration. Tracking systems are in place to document care processes including handling of inpatient admissions and of laboratory, radiology, and other test results. There is an interdisciplinary approach to patient care including arrangements for nutrition, pharmacy, mental health, social, physical and occupational therapy services. Emergency drugs and procedures are in place. Patient wait times are less than 15 minutes (Mackey et al., 1999). The ANC has arranged for 24/7 call. There is a phone triage process in place. Universal precautions are provided for and followed by all clinic staff and faculty.

6. Billing and insurance systems. The clinic possesses or uses a billing system that is accurate, timely, cost effective, efficient, and customer friendly as evidenced by satisfaction surveys. The ANC accepts third-party insurance payers as appropriate for that particular center's patient population.

7. Administration and governance. Policies and procedures are established, documented, and regularly updated. Outside consultants are used for process improvement efforts and audits. The ANC has a long-range plan that is regularly updated. Staff meetings are held regularly with agenda input from all staff and faculty. The ANC posts and adheres to a statement of patient rights. There is a balanced interface and support among the ANC operations, the parent academic unit, and university administrative departments.

8. Marketing efforts. There is a detailed marketing plan with a

budget and staff resources to support the marketing efforts. Outcomes of the marketing efforts are tracked, and the plan is updated regularly.

9. Clinical records and information systems. The systems are comprehensive (medication list, flow sheets, allergies, problem list, SOAP format, confidentiality issues, health maintenance), retrievable, and achievable. These systems are integrated and used by all ANC faculty and staff. Appropriate backup systems are in place.

10. Credentialing and continuing education efforts for faculty and staff. Personnel records verify current licensures and certifications. The center has adequate library references. All faculty and staff maintain and document current CPR certification. Continuing education is encouraged through available funding and release time. Credentialing and certifications of laboratory and radiology services used are verified and current. The ANC is in compliance with CLIA regulations and other appropriate regulations (for example, pharmacy services).

11. Facilities and environment. Facilities and environment should be conducive to patient convenience (for example, hours of operation, patient parking); regulatory adherence (for example, fire inspections, ADA compliance, emergency evacuation plan, personal protective equipment and medical safety data sheets in compliance with OSHA standards); and efficient operations should be maintained (for example, appropriate and up-to-date diagnostic, treatment, and electronic equipment; and routine equipment inspections).

12. Health education and wellness services. Personnel should be trained in health education and wellness type services. A documented program plan should include a needs assessment for targeted populations. There should be personnel training for the plan's implementation. Documentation of these services should be found in patient charts.

13. Patient/corporate satisfaction feedback. A regular feedback mechanism is in place as well a mechanism to receive and act on complaints.

14. Faculty and staff issues. Pre-employment (for example, security checks, immunizations, health screenings) and orientation plans (for example, team approach to patient care, clinic policies and procedures, job duties, expectations) should be implemented. Employee satisfaction (for example, benefits, retention rates) should be monitored and issues addressed. Where appropriate, nurse practitioners should have faculty status and the school of nursing should employ the staff. Faculty and staff have security clearance before starting employment.

Summary

ANCs, by definition, deliver more than clinical services to patients and communities. The unique identifier that separates ANCs from other primary care/ambulatory care centers is the educational service they offer to students and other faculty in the school of nursing to which they belong. Therefore, measuring the quality of an ANC must include a measurement of the educational properties that it possesses in addition to the usual quality measurements by such organizations as AAAHC, JCAHO, CHAP, and AACN. Unless these properties are included and measured by an ANC, it becomes difficult to justify the existence of such a clinic within a school of nursing.

ANCs are encouraged to develop quality evaluation programs aimed at evaluating the educational aspects as well as the administrative and clinical aspects of their operations.

Table 1.

Common Survey Areas of Major Accrediting Agencies *

Administrative Activities

Administration

Facilities and Environment

Clinical Activities

Records

Diagnostic Imaging

Governance
Managed Care Professional
Delivery Organization
Patient Rights

Professional Improvement
Quality Management
Quality of Care Provided
Teaching and Publication Activities

Emergency Services
Health Education and Wellness
Immediate/Urgent Care
Occupational Health
Overnight Care
Pathology and Laboratories
Pharmaceutical
Research Activities

- * Accreditation Association for Ambulatory Health Care Guidelines
- * Joint Commission on the Accreditation of Healthcare Organizations Guidelines
- * Community Health Accreditation Program Guidelines

* American Academy of Ambulatory Care Nursing Standards

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ABSTRACT: This article is a critical analysis of the American Health Security Act of 1993. Although AHSA was soundly defeated when first proposed, parts of it have been enacted into law in 1996, with the prospect of further piece-meal enactments in the future. It includes matters of fundamental importance to American mental health practitioners, to vulnerable citizens with psychiatric disorders, to their families, and to their few champions in medicine and law. Utilitarianism is the unstated philosophical substructure of AHSA and its legislative progeny, i.e., whatever cuts medical costs and saves money is good.

The author delineates AHSA's mental health entitlements and limitations of in-patient, out-patient, and other patient care. She enumerates a dozen major imperfections and dangers of this mental health law, especially its medical utilitarianism emphasizing outcomes and quality of life. Dr. Cosman argues that medical cost, outcome, quality of life, and managed competition threaten the essential liberties and the lives of older persons, persons who are chronically ill, fatally ill, and most particularly those who are mentally impaired. She concludes that if limited

money, medicine and time are invested only in inevitable medical success, then America's medicine by its medical law will be Medical Darwinism encouraging survival of the fittest by requiring extinction of the unfit.

When was the last time you read the failed Clinton Health Plan of 1993 called The American Health Security Act (AHSA)?(1) AHSA's 1,364 pages of legislation include sections of critical importance to American mental health practitioners, to the vulnerable citizenry with psychiatric disorders, to their families, and to their few champions in medicine and in law. Everyone concerned with America's mental health care should read AHSA. It is astonishing psychiatric cultural history. Analysis of AHSA, however, is more important for providing context for medical legislation directly derived from it and already law of the land.(2) Furthermore, studying AHSA now provides warnings of other AHSA sections proposed for forthcoming medical laws. Utilitarianism is the unstated philosophical substructure of AHSA and its legislative progeny. What is good for the state is good for the patient. Whatever cuts medical costs and saves medical money is good. A patient under capitation payments is simply one head among many hungry heads. Individualism, integrity, and autonomy are out-worn ideas no longer valid for modern America where medical costs are stratospheric, and chronically ill patients live expensively long lives with poor medical "outcomes," and dismal "quality of life." Every American must have equal access to medicine. Those who get too little must get more. Those who get too much must get much less.

Many powerful people lamented AHSA's failure to become American law, especially its mental health provisions. Some politicians still bemoan it as a lost necessity.(3) Other legislators are determined to implement AHSA slowly, surreptitiously, and part by part. Whole sections of the Clinton health plan already have been incorporated essentially unchanged in legislation Congress passed into law in 1996. For instance, the Health Insurance Portability and Accountability Act (HIPAA, familiarly called Kennedy-Kassebaum) contains approximately one hundred pages directly from AHSA of criminal penalties for doctors who minimally, even unintentionally, violate the law.(4) Other AHSA restrictions governing Medicare patients were interpolated into Section 4507 of the Balanced Budget Act of 1997.(5) Moreover, Senator Hillary Rodham Clinton and her adherents know that AHSA is not dead but hibernating, ready for vigorous reawakening, hungry and rapacious, whenever the political season warms and time converges with her opportunity

Dangers to liberty, restrictions of choice, and noxious naivete of AHSA are most dramatic in section 1115 defining Mental Health and Substance Abuse Services.(6) Note that section title. That itself should sound an alarm. Throughout the legislation, mental illness and drug addiction are joined as equals: as if their origins, expressions, requirements, social symptoms, pariah status, and remedies are identical. Moreover, legislation provides mental health care to all Americans, increasing manyfold the citizenry entitled to free or quasi-free psychiatric and or mental health care. Entitlements suddenly open to those who do not need them. Banal and ordinary treatment is provided to those who already have superior treatment plans.(7) Those with will and money to pay for better are required to accept worse care. Lamentably, the poorest, sickest needing most help have severe restrictions on their chances for getting it.(8)

For the mental health community, AHSA and its progeny in the law are obnoxious perversions of an attractive chance to do good. Good intentions, however, can lead to worse evils than sinister intentions. Malignant neglect can murder as effectually as guns or poisons. As the last half century of mental health legislation and litigation in America and in Germany suggests,(9) though we may be alert to obvious wickedness and readily fight it, we easily can be lulled into unwittingly killing precious excellence, suspending moral abhorrence, and encouraging the unspeakable to perform the unthinkable.

Permeating the mental health provisions of AHSA is the Hegelian philosophy that whatever is useful for the country is right for the person. AHSA's mental health sections demonstrate the moral irresponsibility of promising ideally generous benefits whose astronomical, reason-defying costs can be contained only by the cruelest restrictions not only upon those who cannot pay but upon those competent to buy care but who by law are forbidden to obtain it. I repeat: AHSA requires that every American have no more and no less mental health care than any American. People currently thriving with their own private mental health care would be prevented from getting it, even when ready, willing, and able to buy it with their own money. People accustomed to selecting among options their good insurances provide would be forced to take whatever the universal plan elected to provide. Independence, personal autonomy, and integrity of physician and patient choices not only are not permitted but are punished. Since most law guiding and guarding medicine in our nation is no longer civil law but criminal law, the punishments for violating medical law include prison.(10)

Mental health provisions in AHSA are defiant attempts at social engineering. If you have not studied these recently, I will delineate AHSA's mental health entitlements and limitations of in-patient, out-patient, and other patient care. Then I shall list a dozen major imperfections and dangers of this mental health law, especially its medical utilitarianism emphasizing outcomes and quality of life.

Readers familiar with Hegel and medical utilitarianism in prewar Germany will find the mental health sections of AHSA terrifyingly suggestive of psychiatric Darwinism wherein survival of the fittest requires extinction of the unfit.(11) Those of us studying law and old enough to remember utilitarian controlling ideas and their perversions, know well that judging who shall live and who shall die via AHSA'S criteria of cost, outcome, quality of life, and managed competition will pose great inconveniences upon the liberty of the young and healthy. But medical cost, outcome, quality of life, and managed competition threaten the essential liberties and the lives of older persons, persons who are chronically ill, fatally ill, and most particularly those who are mentally impaired. Their costs of care are astronomical, their outcomes are gloomy, and their life quality to those who observe them, not necessarily in the patients' judgment, is life not worth living.

I believe with Justice Louis Brandeis, that our enemies often are our friends innocently trying to help us.(12) But worse are those who posing as our friends pridefully insist they know our minds and our bodies better than we do, and insist upon controlling what is done and not done to our minds and our bodies. The greatest dangers to liberty lurk in the insidious encroachment of people of zeal, well-meaning but without understanding.

Mental Health Provisions in the Clinton Health Plan

Of the twelve titles in the American Health Security Act the most important for mental health care are the first(13) and third.(14) The full Act's twelve titles are:

1. Health Care Security
2. New Benefits
3. Public Health Initiatives
4. Medicare and Medicaid
5. Quality and Consumer Protection
6. Premium Caps, Premium-Based Financing, and Plan Payments
7. Revenue Provisions
8. Health and Health-Related Programs of the Federal Government
9. Aggregate Government Payments
10. Coordination of Medical Portion of Workers' Compensation and Auto Insurance
11. Transitional Insurance Reform
12. Temporary Assessment on Employers with Retiree Health Benefit

Costs

The major mental health provisions are found in Title 1, Health Care Security, subtitle B, Part 2, Description of Items and Services Covered, section 1115, called Mental Health and Substance Abuse Services.(15) Other mental health material is sprinkled throughout the legislation, especially in Title 3, subtitle F, Part 1, on Financial Assistance, section 3501, on Authorization on Public Health Services Initiatives Fund.(16)

Examining Mental Health and Substance Abuse Services repays time and effort. Before I suggest a dozen imperfections and dangers of this mental health law, let's review in detail the three separate though interrelated services of which we all would be beneficiaries: (1) In-patient and residential mental health and substance abuse treatment; (2) Intensive non-resident mental health and substance abuse treatment; and (3) Out-patient mental health and substance abuse treatment. Eligibility under the law is not always what it seems. The eligible mental health patient has, or has had during the one year preceding the date of treatment, a diagnosable mental or substance abuse disorder, and is experiencing or is at significant risk of experiencing functional impairment in family, work, school, or community activities.(17) A person under treatment for a diagnosable disorder but not functionally impaired in family, work, school, or community activities, shall be treated as if so impaired.(18)

Integrating entitlements theoretically is logical and case management is AHSA'S reasonable method. But case management for mental health and substance abuse patients under Section 1115(b)(2) is only conditional assistance to an individual in gaining access to needed medical, social, educational, and other services.(19) An eligible American will not necessarily get what he is eligible for.

Eligibility for case management requires the patient to be receiving out-patient mental health and substance abuse treatment.(20) But a health plan administering AHSA benefits (for instance, your local HMO or my state health plan) has the discretion to offer case management or not.(21) An amusing, unanticipated result of case management might be that Americans without mental health problems nor the luxury of an advocate helping them to "needed medical, social, educational, and other" services shall be required to fend for themselves against gatekeepers glad to keep them away from expensive, necessary services. Therefore those suffering mental health and substance abuse problems shall get better medical care than those without mental impairment. Unlike discretionary case management, other outpatient services such as psychiatric screening, assessment, and crisis services are compulsory.(22) Every health plan must provide under section 1115(b)(3) out-patient mental health and substance abuse treatments and services to all enrolled people. Since enrollment in AHSA is obligatory for every American, then, ipso facto, mental health and substance abuse entitlements of screening, assessment, and crisis service would be open to everyone.(23) Collateral services benefit family members of people receiving mental health and substance abuse treatment, serving the wife, husband, father, mother, sibling of a person with a mental or addictive disorder.(24)

Note well the consistent, inextricable linking of Mental Health to Substance Abuse treatment. Every American is considered a public health client. Jostling for space in the same psychiatric life boat are the alcoholic snoring in a downtown doorway, my wealthy neighbor's child with lithium-managed bipolar disorder, your fiscally competent nephew with minor retardation and severe cerebral palsy, the new mother with postpartum depression, all the city's drug addicts, plus you, and I.

By January 1, 2001, seven years after the plan was to become law, mental health care was to be totally integrated with all other bodily health benefits for a person with a health security card.(25) Parity meant equal poverty of benefits. Before parity, the maximum number of residential mental health care days was thirty days in a state-licensed facility

legally authorized to provide treatment in a least restrictive setting.(26) A dangerousness exception extends the annual aggregate maximum to sixty days if the individual receiving treatment poses a threat to his or her own life or to the life of another.(27) AHSA gives no clue to what happens after the patient hospitalized for two months is not yet cured, a bureaucratic finality few serious mental conditions obey. Anyone not well in sixty days is out on the street. Inpatient hospitalization for substance abuse treatment covers only medical detoxification associated with withdrawal from alcohol or drugs.(28)

Intensive non-residential treatment, the second form of entitlement, offers diagnostics or therapeutics in non-residential hospitals, day treatment centers, psychological rehabilitation programs, ambulatory detoxification programs, home-based mental health services, or behavioral aid services.(29) But intensive non-resident coverage is discretionary to a health plan, which for philosophical or fiscal reasons may or may not offer it. Intensive non-residential treatment should avert the need for the more expensive residential, in-patient services. Annual limit to intensive nonresidential treatment is an aggregate 120 days.(30) Two days of intensive non-residential treatment equal one day of in-patient treatment.(31)

Therefore, a patient could exchange the maximum annual in-patient ration of sixty days to obtain twice as many days of care via this intermediate method between in-patient and out-patient. Intensive non-residential treatment, if available at all, can be extended an additional sixty days if medically necessary.(32)

The third mental health entitlement is out-patient treatment.(33) Nine classes of out-patient activities under section 1115(e) include: (1) Screening and Assessment; (2) Diagnosis; (3) Medical Management; (4) Substance Abuse Counseling and Relapse Prevention; (5) Crisis Services; (6) Somatic Treatment Services; (7) Psychotherapy; (8) Case Management; and (9) Collateral Services.(34)

Limitations on out-patient psychotherapy and collateral services (for patients' families) are thirty visits each.(35) Further out-patient privileges, available at the discretion of the health plan, might prevent hospitalization or facilitate earlier hospital release. Out-patient day exchanges with in-patient day limits are four-to-one.(36) For substance abuse, the four-to-one exchange is at the discretion of the health plan. In order to prevent relapse, thirty extra days of group therapy are available to an addict.(37) But a second in-patient detoxification period, if the first has failed, will be available only if the health plan determines that there is "substantial chance of success."(38)

Cost sharing is critical to the proposed mental health services.(39) Americans either would be obliged to share costs by co-payment, out of their pockets, or coinsurance, presupposing they possess it. For in-patient care, coinsurance is 20% of the applicable rate.(40) Cost sharing in intensive non-residential care requires no co-payment but a 20% applicable co-insurance payment. For out-patient services, cost sharing is \$10.00 per visit as co-payment and 20% applicable coinsurance.(41) Psychotherapy has different cost sharing arrangements. Co-payment out of pocket is \$25.00 per visit (until the then expected year 2001) and thereafter \$10.00 per visit.(42) Co-insurance is 50% of the applicable payment rate (to the then expected 2001), thereafter 20%. (43)

AHSA's mental health benefits also are extended to unknown patients yet to be discovered. AHSA stresses bringing mental health and substance abuse services to the patients, not waiting for patients to ask. Progressively larger amounts of money are budgeted for patient transportation, community and patient outreach, patient education, foreign language translation services, and such other services as the Health and Human Services Secretary deems appropriate.(44) Title III, in the financial assistance section of subtitle F, originally mandated \$100 million for

fiscal year 1995, \$150 million for fiscal year 1996, and then \$250 million for each of the years between 1997 and the year 2000.(45)

Remarkably, increasing the number of clients is the proposed method for reducing total costs of universal medical care. Extensive, expensive Outreach Services are thought necessary prelude to integrating mental health and substance abuse services into the comprehensive benefit package.(46) Therefore AHSA requires reports.(47) Under section 3511(b) (3), a report must provide information on the extent to which each health provider furnishing mental health and substance abuse services participates in one or more regional or corporate alliance health plan, and, in the case of providers not participating, why not.(48) The report also must state the amount of money providers get from health plans.(49) Then, under section 3511(b) (8), the report must identify the changes in participation "and certification requirements needed to achieve integration of programs and providers into Health Plans."(50) This is not innocuous fact-collecting. Mandatory reportage has ominous implications under PORTS, the "Patient Outcomes Research Teams," and their annual ORPs, or "Outcome Performance Reports." Practitioners who do not conform are ousted from medicine. It already is happening.

AHSA also endangers confidentiality. It offers supplemental formula grants to states to provide money for coordinating and monitoring, especially (1) management information systems and (2) establishing linkages between providers of mental health and substance abuse services, primary care providers, and the health plans. Under the information infrastructure proposed in AHSA, confidentiality in psychiatry would vanish.(51) Today, under HIPAA, failure to provide medical record data is punishable by five years in prison.(52) Both physician and patient are liable under the law for not providing information or for providing "false" or incomplete information. Presumably, under current law, HIPAA, a psychiatrist and patient unwilling to report an episode or document a session could share adjoining cells.

Who plans for whom? Government plans for everyone in determining services: (1) from the most restrictive in-patient to medium restrictive intensive nonresidential to least restrictive out-patient; (2) from the most expensive in-patient to the medium expense of intensive non-residential to the least expensive out-patient psychotherapy or other services; and (3) from the most time-intensive twenty-four hour monitoring of in-patient treatment to the medium intensity of intensive non-residential treatment during the day to the least intensive partial day, hourly, or partial hour out-patient mental health activity. Levels of restriction, expense, and intensity require exchanges because of inevitable money demands. Intermediate intensive non-residential treatment is exchangeable two-to-one for in-patient. Outpatient is exchangeable four-to-one for in-patient days for the annual health care ration per American.

Any seductive logic to AHSA's three mental health and substance abuse services slips into oblivion when costs are counted. A health plan not able to afford or not willing to afford certain services is not obligated to provide them. Money determines whether there is or is not a basic triad of in-patient, intensive non-resident, and out-patient mental health and substance abuse treatments or simply the banal old pair of in-patient and out-patient care. Coercion would determine who gets how much of which service. Everyone in America is part of the who gets what. No matter how intelligent, independent, or wealthy, no one can have more and everyone shall have less mental health care. Rationing per head is only one of a dozen perils of the plan.

Imperfections and Dangers of AHSA's Mental Health Provisions

Naming

Mental health and substance abuse services are inextricably yoked in AHSA. Eligibility is identical, definitions of services, and limitations

are the same for the one and the other. While thousands of Americans have dual diagnosis of mental disorder and addiction to either alcohol or drugs, mental illness and addiction are not equivalent. While substance abuse may be a sub-set of mental ill-health, mental disease is not necessarily consequent upon substance abuse. Nor is mental disability a sub-set of addiction.

Linguistic separation is essential. Mental disorders, subject to their own prejudices and stigmata, must avoid customary imputation of moral fault and free will attributable to addiction. One can elect to snort cocaine but I know no free choice for trying on an episode or two of schizophrenia. If genetic predispositions exist for alcohol and psychotropic drugs, and if chromosome studies ultimately prove heroin and crack addictive only to people with particular genetic configurations, nevertheless addiction almost always has a volitional beginning. Despite neurochemical and biochemical characteristics of drug dependency, free choice usually governs experimenting with drugs or using addictive substances for recreation or for escape. Volition in initiating addiction incites social censure. To attribute choice and free will to depression, neurosis, and psychosis is ridiculous and dangerous.

Linguistically linking addiction with mental illness also adds unjustifiable social stigma of crime. Criminal acts, often concurrent with addicts paying for their drugs, such as assault, burglary, theft, and murder, by unjust implication attach to mental dysfunction, tainting mental disease with the stench of criminality. Inappropriate dual classification also places the mentally ill and mentally disabled at unnecessary risk to lose benefits when political power reduces them, as is inevitable, and as is demonstrated vividly in Canada and England.

Any classifying term or title either over-inclusive or under-inclusive is dangerous in an entitlement program. Classes incongruently joining conditions, as mental health and substance abuse, either inappropriately add those who should not obtain particular benefits or exclude those who should. Where there is no dual diagnosis nor overlapping of causation, there still may be some similarity in expression of bizarre behaviors, tendency to relapse or recidivate, and favorable response to similar or identical drugs, psychotherapeutic techniques, and behavioral therapy. Nevertheless, the person with depressive, neurotic, or psychotic disease has enough problems handling reality without a credulous public(sections) nomenclature-induced phantom distortions.

The most dangerous practical effect of equalizing mental illness with substance abuse is competition for money. Historically in America both services have been under-funded.(53) AHSA requires them to compete for the same limited funds. Moreover, mental health money now is being spent inordinately frequently in prisons.(54) Consider California's mental health expenditures. In 1994, Dr. Aretha Crowell, Director, Los Angeles County Department of Mental Health, correctly warned that health reform must provide appropriate treatment and rehabilitation for the mentally ill or society will pay directly via emergency rooms, jails, courts, group homes, or other expensive institutional, responses, including societal costs of urban violence and homelessness.(55)

Los Angeles is notorious for having more mentally ill in jails than in hospitals.(56) I intentionally use figures here from testimonies favoring AHSA's introduction in 1994. Nearly 10,000 people treated in jail programs for the mentally ill cost state government \$5,000,000 in 1992 through 1993. According to Dr. Crowell, about 75% to 80% of people seen in psychiatric emergency rooms in Los Angeles have a substance abuse problem. "Bizarre behavior brings them to us, but we find that one-third have only a substance abuse problem, the others having a dual diagnosis of mental illness and substance abuse. We estimate those with only a substance abuse problem cost the psychiatric emergency rooms about \$3,000,000. Law enforcement costs are an additional local burden of these untreated

illnesses."(57)

Fortunately, public health statistics true for one large California city especially burdened with urban blight, unemployment, high drug use, and large numbers of illegal immigrants are not the psychiatry-addiction-unity statistics of the nation. Legislators generalizing from Los Angeles' mental health statistics to all America are as wrong as those who see cardboard sheds and shacks of the homeless defining American architecture, or dinners of poverty-dwellers exemplifying American cuisine. Mental illness must be disassociated from substance abuse.

Swapping and Limiting Use

The thirty day annual limit on in-patient care, with the additional thirty day extension in case of dangerousness to self or others,(58) and the thirty visit limitation on ambulatory psychotherapy care,(59) are familiar to current health insurance and unremarkable. But they are retrogressive in meeting the purposes of AHSA, to prevent catastrophic illness costs and to promoting universal access to health care.(60) Imposing limits on coverage will not control costs. Costs can be controlled without the day or use limitations.(61) Swapping psychotherapy sessions for in-patient days causes several problems. First is the conceptual difficulty: if a basic purpose of health reform is protection to families against financial ruin because of catastrophic illness, then, trading away in-patient coverage for out-patient psychotherapy will leave most vulnerable those who need the most help.

Second, swapping probably will raise not lower costs because relatively few people use in-patient care relative to psychotherapy.(62) Yet the pool of people likely to trade away in-patient days is relatively large. No strong research evidence suggests that psychotherapy is an important substitute for in-patient care. "The drafters of this provision appear to have lost sight of the fact that insurance benefit is not intended to provide a pot of money to use but is instead intended to offer financial protection against the consequences of illness. In general, health insurance should be something you do not want to use."(63)

Enticing troubled people to swap in-patient days for either the two-for-one intensive non-residential services or for the four-for-one out-patient psychiatric visits is likely to deprive the people who most need the most restrictive in-patient care at their times of most dramatic crisis. Either such people will be forced into whatever is left of a public sector mental health system, if such residue will exist, or they shall wander about America's streets untreated.(64)

Patient discretion to swap in-patient treatment days for out-patient treatment sessions must be counterbalanced by a mechanism to avoid both excessive utilization by the healthy and routine denial of in-patient care to the most desperate and the most dangerous.

Serious Chronic Mental Illness

Current Medicaid benefits for people with serious, long-term mental illnesses are far more generous (and fiscally more expensive) than AHSA provides.(65) Reducing benefits to already impoverished and troubled citizens will exacerbate not fix current community problems. AHSA's mental health benefits are ordinary, traditional, and should be appropriately integrated with, not substituted for, Medicaid benefits. To make case management and intensive non-residential therapy discretionary, and then to ration the amount to thirty days annual cumulative service, with an extended increase to sixty days only if "medically necessary," is simply inadequate for our most seriously mentally distressed population currently under expensive but nevertheless reasonably efficacious treatment.

Treating serious chronic mental illness is stunningly costly. In California, for example, seriously mentally ill adults are now being treated under capitation plans at \$17,000 per person per year versus the cost of such a person to the state of \$100,000 annually in a state hospital

or \$60,000 per year in a skilled nursing home.(66) Los Angeles County has shifted 500 most costly clients, averaging \$30,000 per person per year to Integrated Services Agencies costing merely \$17,000 to \$20,000 annually. Appropriate field interventions reduce emergency room and jail episodes for these seriously mentally ill. Such nontraditional interventions, however, are not provided for in the pedestrian, traditional, anti-innovative AHSA mental health benefits.

AHSA resembles Medicare's generous need-anonymous entitlements gratefully abused by the middle-class and the wealthy elderly.(67) They do not know their peril in exchanging their true freedom for nearly free medical care. Nevertheless, people profiting most from AHSA's mental health advantages are those who need them least, namely: the mildly mentally ill middle class and wealthy people of all ages now paying via insurance or their pockets.

Banal Benefits in Mental Health Managed Competition

While states such as California, Oregon, and Arizona have experimented with integrated programs that are both cost-effective and medically-effective,(68) the Clinton AHSA benefit uses hospital bed days as the fulcrum around which the whole program pivots. With millions of middle class and wealthy people suddenly brought into a single mental health system which heretofore has studiously distinguished between the public and the private, and with 37,000,000 new patients from America's currently "uninsured," disproportionate numbers among the unemployed, under-employed, and homeless, this emphasis upon the old inpatient treatment is sure to lead to financial disaster.

Almost everyone in health law uses the phrase "managed competition." But even its originator, Dr. Alain Enthoven of Stanford, has never explained it efficiently or memorably.(69) When I am on a lecture podium handling questions, I attempt a requisite definition, but I do not truly believe it. Enthoven's impassioned rhetoric and slippery definitions cover such statements as "naked managed care is not a panacea but modified managed care may be."(70) Whatever that might mean, managed competition of mental health care does not yet exist. Like the coming Bodhisattva of the Buddhists, it has some avatars, preliminary earthly impersonations of blessedness presaging the perfection to come, namely in some competitive HMO mental health facilities and in some intelligently integrated psychiatric and social services nationwide.

Managed competition in its current inchoate forms seems to work best for acute mental illnesses with brief and immediate treatments.(71) But chronic mental illness cannot so simply be cured or its symptoms ameliorated or remitted. DRGs, the "Diagnostic Related Groups," may function as cost-controls in hospitals reasonably well for many purely physiological ailments such as infected gall bladders and comminuted tibial fractures. DRGs and managed care methods do not work nearly so well for serious mental disorders.

Even avid Clintonians recognize this. In 1993 the Little Rock Working Group on Mental and Substance Abuse Disorders in Health Care Reform, acknowledged inadequacies of managed competition for mental health:

Exactly how chronic care problems will be accommodated in a managed

competition model has been the subject of some debate. Because the majority

of health care expenditures are attributable to individuals with chronic illness, the Working Group believes that ... (for) persons with chronic illnesses including mental and substance abuse disorders and long term treatments ... the quality of care will not improve for this population

and

costs will not be controllable.... Individuals with severe and persistent

mental and substance abuse disorders often require some form of care over a

long period of time, including services for acute episodes, maintenance and rehabilitative care, and a range of services that do not fit a classic

'acute care medical model.' ... An expanded form of managed competition may

be required to more adequately respond to the long term care needs of this

population.(72)

Strip away the sycophantic and sociological jargon. This pro-Clinton Little Rock Working Group correctly undermines the total concept of AHSA: Capitation alone will lead to undertreatment of chronically ill

populations. According to Enthoven, unmodified managed competition does not

work for all people. Mechanisms must be devised to provide adequate funding

to Accountable Health Plans to permit enrollment and to minimize the risk

of responding with inappropriate or inadequate care.(73)

Some health plans not only will be penalized but probably bankrupted by patients with severe, persistent mental health and substance abuse disorders. Therefore the Group suggests types of risk-adjusted premiums, mixtures of capitation and fee-for-service systems.(74) AHSA alone does not intelligently handle the problems of refractory mental health diseases and chronic psychiatric problems.

Co-Payments

Requiring 50% co-payments for out-patient services probably derives from reasonable desire to limit abuse of mental health services by those who do not need the services in the first place; however, the co-payments are retrogressive. Legislation requires that people not count out of pocket costs spent on alternatives to inpatient care that in other aspects of the Plan would count towards a maximum out of pocket liability.(75) That has the strange effect of vitiating a major purpose of mental health plan coverage. Ostensibly one reason for the total overhaul of American medicine is to provide against catastrophic costs.(76) Requirements that people pay co-payments, but not count them, violates this ostensible protection against catastrophic costs.

The alternatives to in-patient care are only used by individuals with severe mental health/substance abuse problems. These individuals with a high degree of certainty incur ruinous levels of expense. Exempting their

out of pocket costs on the alternatives from the maximum liability rule presents a 'pure transfer' from one of the sickest segments of the

population to the rest of society. This is hardly in keeping with the humane and efficient vision that the President has set out for the treatment of mental health/substance abuse problems in America.(77)

Discretionary Alternatives to In-Patient Care

Unfortunately each health plan has the discretion to provide or not to provide alternatives to residential and in-patient mental health and substance abuse care. Health plans therefore easily will compete on the basis of cleverly selecting risks. Plans not offering the intermediate intensive nonresidential treatments clearly aimed at those who need them most, namely those with severe mental health or substance abuse problems, will not attract them as volunteer subscribers and will not have them allocated to them if, as is likely, many of these people do not sign up for alliances or health plans at all. Health plans not enrolling the severely ill will make more money and more profit than those which do. Insurance companies currently mining the risk pool for the healthiest diamonds tend to cede away those who are riskier. Therefore, when yearly report cards are granted, such clever health plans will look grand on comparative physical scales not because of inherent medical excellence or efficiency but clever avoidance of risky characters.

Health plans also were expected to be able to fully integrate mental health and substance abuse care into the total health system by the year 2001. Those plans persistently and directly avoiding the risks would not have the requisite experience either to manage or to deliver mental health care.

Case Management

Like the intermediate non-residential intensive treatment, case management also is at the discretion of the health plan.(78) If case management is as important as it appears to be as an integrating force reducing duplication, appropriately joining physical and mental therapies, and providing those ancillary educational and other benefits to prevent relapse and recidivism, then only those health plans offering case management will attract patients who require it. Therefore, like intensive nonresidential treatment, adverse selection will overburden those health plans offering case management, a logical integrated function. Adverse selection will free other plans from substantial numbers of the mentally ill or the addicts. Therefore, their yearly finances will look better and actually be fiscally sounder.

The opposite result also is possible. Those plans not offering case management, like those not offering the intermediate intensive non-residential treatment, might incur higher costs than those plans which do, even for their more limited number of patients. Even with clever risk selection, there are such vast numbers of people needing or wanting mental health and substance abuse care that modest or even small numbers of those treated with expensive care such as in-patient services will ultimately prove more expensive to their plans than the hordes of enthusiasts or assignees to those plans offering case management and intensive non-residential treatment.

Long Term Care

No where in the mental health section is long term care specifically addressed.(79) Perhaps, AHSA drafters assumed that the most serious mental health or substance abuse problems already are handled under their section specifically dedicated to long term care. They are not. Everywhere that mental health and substance abuse care is mentioned specifically, it is treated separately because mental health and substance abuse coverages were expected to be slowly integrated into the plan (over a proposed seven years culminating in parity in 2001). Long term care, probably the most expensive of all mental health care, does not necessarily have to be inpatient and can, one way or another, be community-based. As AHSA suggests, long term care may include personal assistance services for those three of five

activities of daily living impaired, such as feeding, clothing, and toileting.(80)

Long-term care entitlements, however, appear aimed at the aged. Available money for long term care under AHSA depends upon what percentage of the population is over seventy-five years of age. What about those suffering from such chronic cataclysms as birth defects? Are certain chronic and irretrievable mental health problems such as retardation included here? What about serious schizophrenia, bipolar disease, and deep depression which simply will not respond in the mandated thirty or sixty inpatient or even the 120 days of intensive non-residential AHSA allotment?(81) And what if people living longer increase the population of over-seventy-five-ers to an "unacceptable" number? Who will make the choice between treating Grandpa James with Alzheimer's versus young Susanna with autism?

Appropriate psychiatric treatment can reduce relapse rates in schizophrenia from 80% to just over 20%, in bipolar disorder from over 80% to approximately 30%, and in major depressive disorder from 70% to under 20%. (82) So Frank Docherty, M.D., Clinical Professor of Psychiatry at Tufts, testified on November 8, 1993.

We are in the center of a wave in new knowledge that has developed over the

last two decades in both basic neuroscience and psychological sciences that

has the promise to usher in an entirely new era in the effective treatment

of the mentally ill. All of this will be lost without the acceptance of our

societal responsibility to fully and completely care for all of our mentally ill. (83)

Though Dr. Docherty correctly rails against the unrealistic lifetime limits that insurance companies set for mental health coverage, emphasizing actuarial rather than clinical data, nevertheless AHSA uses the very same limits. (84) AHSA's time and condition limitations, like current insurance companies, will force patients into the residual public sector, if it exists, thereby overburdening that system.

Children and Adolescents

Limitations on access to care in the current system have shifted responsibility and cost to public mental health and substance abuse systems. While critics of current health insurance and advocates of the Clinton Health Plan damn the current system, they do not appreciably improve it in the proposed overhaul of American medicine. For children and adolescents both burden and cost have been shifted to the systems controlling child welfare, education, and juvenile justice. (85)

As Dr. Bernard Arons, Director, Center for Mental Health Services, Substance Abuse and Mental Health Administration, Public Health Services, Department of Health and Human Services maintains, "Even worse, sometimes these overburdened systems are not able to provide needed services, and the individual goes without treatment. Eventually, we all bear the cost of delays or gaps in service provision." (86) He maintains that a comprehensive array of services and flexibility to provide these upon medical necessity "produces better outcomes than those experienced with traditional benefits. There is also evidence that capitated approaches result in less costly provision of health care services." (87)

Dr. Arons' glittering generalizations in his writings and speech call for full integration of the mental health and substance abuse treatment system uniting currently uncoordinated federal, state, local, and private

organizations, general hospitals, specialty institutions, clinics, and office-based clinicians, and integration of mental health with general health care entitlements.(88) He maintains that "creative use of utilization management and reimbursement systems (prospective payment, capitation, prospective budgets, and performance contracts) opens up opportunities for eventually providing deeper coverage."(89) He quotes Richard G. Frank, H.H. Goldman, and T. G. McGuire,(90) in his essay entitled "Mental Health and Substance Abuse Coverage under Health Reform."(91)

AHSA makes no distinction between problems of adults and children. As Senator Wellstone of Minnesota said, "our new national Health Care System must be a revolution in how we take care of our children."(92) Both Senator Wellstone and Oregon's Representative Mike Kopoloski reminded Congress that approximately 14 million children suffer a diagnosable mental illness.(93)

Yet research tells us that some severe problems show improvement in 26 t
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52 sessions, generally beyond the Administration's 30 session limit. Why should the design of our mental health benefit force the decision to hospitalize a child, removing her from her family, from her support system,

just to use a health plan benefit? When hospitalization is the treatment of

choice, it should be available and covered, but we don't want to force that

choice just because that is the only mental health benefit left in the family's coverage.(94)

Actuarial Equivalence

Many people now have far superior mental health benefits than those proposed in AHSA. Large employers, such as automobile manufacturers, and some union plans are considerably more gracious to their subscribers than AHSA.(95) Employees of these companies or union members would suffer diminished benefits if they were required to accept benefits through a regional alliance or their own company elected to emulate local regional alliances. Senator Robert Kennedy long ago suggested a solution to this dilemma of creating a mandatory entitlement for all while depriving some workers of a current excellence. He had included in earlier versions of his mandatory health insurance program the idea of actuarial equivalence.

Current medical benefits actuarially equivalent or better than the Clintonian AHSA benefit package should be permitted to continue. It is wasteful to cut benefits, restructure a comprehensive, generous plan into a Spartan basic plan and then add yet another supplemental plan to attempt to bring benefits back to their former heights. Since AHSA limits the numbers of days and visits, almost no supplemental coverage could span the gulf between the stingy and the grand.

Actuarial equivalence, however, speaks to the problem of AHSA's total effect. Most Americans are reasonably happy with their current coverage. Everyone's autonomy, individuality, and personal control over decisions about their bodies and minds would diminish under AHSA. Actuarial equivalence acknowledges that one size does not fit all, that not all American people need the same things nor want them. Freedom to choose is simpler and more logical than actuarial equivalence. The Federal Employee Benefit Program thrives because of its excellent choice, and its dependence upon consumer not customer satisfaction. Legislators willingly impose on

all Americans restrictions they would not tolerate upon themselves.

Decision-Making Personnel

Most discretionary judgments of who shall get what extension of benefit are made by health plan employees. Who are they? Providers come in many sizes, shapes, and colors, but they do not necessarily make the decisions. "Clerks" decide. Medicare, Medicaid, and insurance carriers employ "clerks" who make discretionary determinations of whether physicians and surgeons shall or shall not provide services to patients. Who are these people who determine who shall receive the life-enhancing medicine or treatment and who shall not? Who selects these decisions makers? What are their academic and practical qualifications? What knowledge do they possess of the individual patient whose mind and body is the subject of the decisions? How can the understanding and discretion of a person who never has seen nor examined nor interviewed the patient, and who is likely to be medically far less educated than a physician, be superior to the judgment of the physician and the patient?

Doctors now are free to treat for free (although treating for free under Medicare can be interpreted as a false claim, a crime against the government) if their intended treatment plans are not approved by third-party payers. Under AHSA physicians are forbidden to countermand prohibitions against care. If care is not medically necessary, the physician providing it and the patient requesting it are guilty of crime. Medical crime, including the crime of providing and billing for "medically unnecessary" medicine and surgery, is punishable by prison.(96)

Determination of what is "medically necessary" has little to nothing to do with what diagnostics, medicine, and surgery are necessary for the diagnosis, treatment, and cure of an individual patient. Medically necessary care is determined by what the third-party-payer will or will not pay for.

Even free care is tantamount to crime, and to be reported and penalized via fines and even prison for "excessive care." For somatic problems, an inexperienced or stupid decision-maker might cause a patient(sections) death. In psychiatry, the wrong decision on premature release or refusal to treat severely homicidal patients might cause not only the patient's death but deaths of others.

Among AHSA proponents who testified at Washington hearings in 1993 and 1994 were members of the gigantic employee health benefits company providing health plans to a large percentage of Fortune 500 companies called Hewitt Associates.(97) Hewitt speakers note an important caution: it is especially important that services be provided by persons specifically trained and with practical field experience, preferably professionals in social work, psychology, nursing, or medicine who have special training in substance abuse.(98) (Note the emphasis not on mental disability but on addiction, and the order of "providers," with physicians placed last). Though speaking of their experience gathered in the workplace by EAPS, "Employee Assistance Programs," the Hewitt speakers stress success of early intervention. Such access to mental health care potentially is available under the Health Security Act's inclusion of screening, assessment, and crisis services necessarily provided by all health plans. But the likelihood is small that enough excellent mental health professionals will work willingly for meager wages to screen, assess, and intervene. Who will train the cadres of bureaucrats required? Who will pay for that training? Who will pay the screeners, assessors, and interveners? Why? When? How?

Under AHSA, special training in addiction and in mental health must be required of all gatekeepers.(99) That is intolerable responsibility to place upon primary care physicians. Primary care doctors will become the least deeply medically-educated, least intellectually engaged medical personnel because as jacks and jills of all trades they have time and spirit to master none.

Managing Managed Care's Management Costs

Though ostensibly knowledgeable commentators state that AHSA's mental health benefit is managed care, it is not. Nothing in AHSA's mental health and substance abuse legislation particularly is demonstrative of managed care except for oblique references to capitation. Though usually united, they are not synonyms. Capitation, the payment for care per head of American no matter how much care or treatment the head or its body requires certainly is a major feature of managed care. But other payment mechanisms can be integrated into a system still called "managed." As friendly critics Dr. Frank McArdle, Dr. John Mahoney, and Mr. Dale Yamamoto of Hewitt Associates, said:

National health care reform legislation should include a managed mental health benefit alternative along with the indemnity Plan option. The principal reason is that current, state-of-the-art managed mental health plan designs would allow health plans to offer plan participants more generous benefits at a relatively lower cost. And if the proper standard
s

are set for those who manage the care, quality may be improved.(100)

Such startlingly vague benefits justify the gigantic risk of total revamping of medical care in America. The Hewitt speakers are among the few favorable to AHSA who have pointed out the necessity for higher administrative costs because of managed care. Hewitt estimates thirty dollars per covered life per year "under the Managed Care scenario to reflect the costs to run a provider network and provide assessments, referral, ongoing review, and management services."(101)

That ludicrously low amount per person must create a new system. No administrative structure exists nor are personnel trained either to lead or to implement enrollment for millions: collecting premiums, adjusting health plan premiums for risk differences, informing consumers with verified information about each health plan, managing customer services, administering the global budget, claims processing, establishing and implementing medical policy for about 750,000 physicians and millions of other providers, managing the alliances as huge bureaucratic entities required to interact with a government bureaucracy, which, if it continues Medicare's dismal efficiency, requires familiarity with at least 20,000 pages of legislation, statutes, manuals, regulations, and directives, and new administrative instructions issued every five hours.(102)

The most specific management data on mental health and substance abuse in testimony to Congress provided information between four and seven years out of date. Even when current, it was illogical. Hewitt's Dr. McArdle quotes a managed health case study to substantiate assumptions in his analysis, not providing names, locations, nor any means of verification, reporting one company's managed mental health benefit begun in 1987. "In one location, they observed an overall 67% reduction in costs in the first two years of the program. The costs have continued to decline at an average rate of 13% per year."(103) "Similar" first year reductions existed at two other company locations with, again, no verifiable data, company names, nor geographical locales. Admission rates for locations adopting the managed program, Hewitt speakers maintain, decreased by 45%, on average, between 1990 and 1992.(104) Other locations not adopting the managed program increased 17% during the same time. The speakers allowed that upward adjustments in the cost of these plans should be made to reflect new pools of uninsured individuals. Nevertheless, "overall cost relationships maintain the same relationship."(105) Virtually all data in the McArdlé, Mahoney, Yamamoto presentations was four years old in 1993,

and still repeated after the turn of the millennium.

Vagueness, confusion, and contradiction define testimony of experts in AHSA's prolegislation history. But occasionally, truth glistened in a slim shaft of bright light.

As far as quality of care is concerned, we have noted a tendency to define

'Managed Mental Health Care' as care provided through Health Maintenance Organizations (HMOs). We would like to emphasize to you that in our experience, the quality of care provided through a Managed Mental Health program can be superior to the average HMOs capitated approach, HMOs are paid a low flat fee per enrollee, and they 'manage' mental health and substance abuse by adopting a system of benefit design or utilization decisions that may, in effect, deny care; and certain poor outcomes, especially for the more serious or chronic conditions, have been documented

by some recent studies.(106)

HMOs couple utilization monitoring with discounted fee for service payments to doctors.

This allows for more appropriate and better quality treatment. But it costs

more than HMOs typically spend for mental health care. The typical HMO targets a budget of approximately \$5 per member per month for mental health

and substance abuse spending. The actual number reported by InterStudy in

1989 was \$2.69. This level of HMO spending is lower than the cost per covered life in the managed mental health programs we are describing, which

is about twice as high. Thus, the managed mental health programs save money

long term by making more effective use of resources, not by denying care in

the short run.(107)

The best most ethical psychiatrist, psychologist, and mental health practitioner will provide the best care that time and talent allow. But that practitioner must be paid. Of course the practitioner might be a member of a religious order which provides his food, clothing, shelter, books, and entertainment. Or the mental health professional may have a generous spouse providing all creature comforts for the non-income producing doctor. Or the doctor may be independently wealthy, and thanks to a rich papa views medical work as private philanthropy with no recompense necessary. But the majority of mental health practitioners in America are intelligent, devoted, ethical, hardworking breadwinners for their families and themselves. Who is the physician who respects his own professional

time, talent, training, and his integrity who willingly welcomes a mental health patient who pays for care a mere \$2.69 per month?

Which legislator in Congress would dare build into a public minimum hourly wage law for unskilled workers a recompense as low as \$2.69? Who in labor law would encourage for the least competent foreign manual laborers a stated recompense per hour, per "professional encounter," per unit of work at the meager rate of \$2.69?

Under capitation, not every patient comes for care and not everyone comes each month. But mental health patients need care, use care, request care, and demand care. So do their advocates. Drugs are costly, written materials are costly, individual therapy is costly, group therapy is costly, residential care is costly. Why are the best and brightest mental health medical practitioners asked to work for a pittance? How can their therapists' starvation recompense benefit sick patients? Mental health clinicians, like other American doctors under managed care, are given the "carrot" of monetary incentives and year-end bonuses to withhold expensive procedures and medications. Clinicians are threatened with a "stick" of withholding a percentage of his or her own salary until year end when expensive-procedure utilization analysis determines that the physician has been frugal enough in depriving patients of as much care as can reasonably be tolerated.

What are the incentives to our best of best psychiatrists and mental health clinicians? What are the incentives to beleaguered families of the mentally ill? What are the incentives to legislators? Why are "ethicists" now debating medical futility?(108) Why are lawyers and ethicists urging the new "duty to die"? (109)

Medical Darwinism = Survival of the Fittest + Extinction of the Unfit
In some countries of the world, and under some medical programs in America, for example in Arizona and Oregon, patients with a mental disorder, chronic nervous condition, or mental impairment with poor prognosis are given a lower priority for treatment than patients with time-limited conditions and with good prognoses.(110) Conditions with low priority receive no money for treatment. Without money there is no treatment.

What are the next steps? Doctors by law cannot treat. Hospitals by law cannot treat. What happens to people whose conditions are serious but whose care is classed as less valuable because its expense is high and its outcome less favorable than those with higher ratings? Those not worthy of treatment must have mere custodial care. Custodial care, however, also is expensive. What shall be done with precious medical resources for those with conditions with poor current outcome and even worse prognoses? Is not custodial care a useless drain upon limited medical resources better applied to medical conditions with more hopeful prognoses?

Now what? Should "hopeless" cases be sent home to their families? Can families care and cope? Wouldn't it be kinder to the families to relieve them of the burden? If acute care is too expensive and custodial care also burdensome to the state, then caring for hopelessly ill people at home prevents the family giving care from being productive. That is socially expensive in lost wages and productivity Parents sacrificing for a sick child must neglect or abandon care of their well children. That would be a waste of parental effort for no possible gain, a social loss for the healthy children, and for the state.

Triage by Hegel's whatever is efficient is fight, leads inexorably to an unspeakable conclusion. But I will speak it. Would it not be efficient and fight to prevent the hopelessly ill from selfishly using resources better applicable to people likely to be cured and to become productive Americans? If given a choice, wouldn't these people want to avoid burdening their families, their siblings, their state? Wouldn't it be kinder to kill them? Would it not be better for all concerned to kill them?

Consider the logic. How can medical conditions which are incurable,

hopeless, futile, "unqualified for life" be permitted to waste limited medical time, medical effort, and medical money? Aren't such incurable, hopeless, futile people unqualified for life? If their treatment is medically unnecessary, aren't they because of their illnesses also unnecessary?

If to treat them cannot be thought essential, imperative, indispensable, obligatory, or required, how can they be? On the other hand, if such a person's life was indispensable, how could we dispense with it? If the life were imperative, how could we not protect it and preserve it? Doubtlessly those who have no preservation-worthy quality of life we can voluntarily, not obligatorily, desire to treat if we have funds enough, world enough, and time enough.

But unfortunately for them, we do not have world enough and time enough. Death is the only logical conclusion. Likewise, national socialist Germany rationalized extermination of those with hereditary or refractory mental disease.(111)

I do not say we should. I do not say we will. I only say we could. Because we did.

Expendability of people with mental impairment is commonplace in the history of American medical law. Remember the Carrie Buck eugenic sterilization case. In *Buck v. Bell*,(112) Carrie Buck, committed in Virginia to the state "Colony for Epileptics and the Feeble-minded" was sterilized because "three generations of imbeciles are enough."(113) That celebrated statement by Justice Oliver Wendell Holmes pertained to Carrie, an ostensibly retarded daughter of a putatively retarded mother, who gave birth to a presumably retarded daughter.

Under eugenic theory until *Skinner v. Oklahoma ex rel. Williamson*,(114) invalidated a statute authorizing sterilization of certain felons, we did it. Or consider the Willowbrook hepatitis experiments on "useless" children in homes for the retarded on Staten Island, New York.(115) Medical experiments on prison inmates(116) such as the Kaimowitz-prevented psychosurgery research suggest our capacity to formulate and actuate notable horrors.(117)

Prejudice against the "hopelessness" of mental retardation, mental diseases, and mental injury is evident in the current social scientific ideas of quality of life, futility, and emphases upon outcomes. Lethal effects of these three ideas are evident in the current case law pertaining to the right to die.(118) Nevertheless, I applaud the courageous Cruzan family in *Cruzan v. Harmon*.(119) I followed with admiration the adventures of the Quinlan's attempt to release from a ventilator their comatose daughter Karen Ann.(120)

I agree with enthusiasm with most holdings allowing an individual the right to refuse heroic life-extending mechanical treatments.(121) Yet I am appalled by the recurring assertions in court testimony and case holdings that death is preferable to disabled lives "not worth living." The concept of releasing people who want to die is fine. The application is faulty.(122) Case holdings and media reviews of the right to die laws act as insidious encouragement of people with severe disabilities to elect suicide.(123)

Quality of life, outcomes, and futile medical intervention also are important themes in medical malpractice cases on wrongful conception and wrongful birth. Courts view some disabilities as fates worse than deaths, with plaintiffs collecting damages for having to suffer the indignities of impaired life. Dr. Kevorkian and his physician-assisted suicide techniques called obitiatry provide thought-provoking material on the uses and effects of euthanasia for the mentally ill.(124)

Cost consciousness, cost efficiency, and cost savings are the major driving practical forces of AHSA. Cost control is the major perverter of morality Are we as a nation so poor that we must expend our aged and incurable? Shall we protect people with disabilities under the Americans

with Disability Act, and then reduce their numbers by killing them off by withholding their medical treatment? (125) Psychiatric problems and mental health programs are particularly vulnerable in cost versus benefit analysis. If we refuse to treat by national policy, and under threat of criminal penalties we refuse to allow citizens or families to obtain from private sources care prohibited by the government (which malevolent clauses are written into HIPAA and the Balanced Budget Act's section 4507), then there are only two rational treatments available for those who not among America's mental fittest are unfit for medical care: neglect and death.

Determining what is medically necessary, namely what the government or insurance plan will pay for, assumes that medicine or surgery are available for treating a particular ailment yielding amelioration of symptoms, prevention of worse symptoms, healing, or curing. The most magnanimous interpretation of "medically necessary" defines as medically unnecessary those therapies "useless" because the patient's problem is too benign or too malign. At one extreme, home remedies or nature or time's passage will cure. Medicine is superfluous gilding to the gorgeous golden natural process. At the other extreme, medicine is unnecessary if the patient's problem is "fatal" and "hopeless." The person will die before medicine can benefit, and intervention would be futile intrusion. For such benign or malign medical problems, medicine would be frittered in wasteful expense. Diseases not responsive to treatment should not be treated. American money, medicine, and time are limited. America must invest only in medical success.

This is medical Darwinism. This doctrine encourages survival of the fittest and extinction of the unfit. Darwinism is good science in the natural world for describing evolutionary process. But social Darwinism is a perversion of the scientific idea. Battlefield triage, for example, may be reasonable in wartime when the necessary purpose is to save as many fallen warriors who shall survive and arise to fight again. Utilitarian triage in peacetime America's mental hospitals and psychiatric offices inevitably leads to malignant neglect.

AHSA's intended cost-effective research and cost-conscious practice was to replace with new doctors the old-fashioned, outworn physicians and surgeons who had pledged allegiance to their patients by having sworn allegiance to Hippocrates and Maimonides. AHSA and its modern progeny in HIPAA and similar legislation would create new doctors who pledge allegiance to America's global budget.

The most benevolent, efficient, and praiseworthy government cannot protect the patients' best interests simultaneous with state interests. Necessarily, governments are selfish beasts concerned with their own longevity, their own fiscal health, and their own privileges. In government-directed managed competition, managed care, and managed medicine, whatever these concepts really represent, capitation is a dangerous moral wedge. Patients are not individuals but mere "heads" classified by diagnosis. Another danger is "outcomes." Another is the idea of "medically necessary." Another is "quality of life." Inexorably these concepts lead to withholding medical care. Withholding is efficient. Withholding is utilitarian. But if the state decides, not the citizen, withholding is not just. Only accidentally is it humane.

Conclusion

AHSA's dozen dangers in mental health benefits are merely typical of the total Act which vitiates its own major stated intentions, such as preventing catastrophic illnesses from devastating families, and assuring everyone, rich or poor, currently insured or not, access to excellent medical care. AHSA's deficiencies in mental health care include over-inclusive naming uniting mental illness with drug abuse, treatment swapping, inadequate time and care for serious chronic mental illness, banal benefits to all, regressive co-payments, discretionary alternative intensive non-resident care, discretionary case management, unrealistic,

meager requirements for long-term care, non-specific programs for children, actuarial equivalents, insufficient critical decision-making personnel, and naive under-projection of managed care's management costs.

AHSA's ideas are detrimental to the mental health of the nation. That failed legislation and its rejuvenations in current law are dangerous disservices to mental health law and the mental health of Americans. Mentally ill people are particularly vulnerable in cost versus benefit analysis. If by national policy of cost reduction we refuse treatment and by threat of criminal penalties we forbid citizens or their families to buy care privately, which already is the law under HIPAA and the Balanced Budget Act's section 4507, then two rational prescriptions for those not mentally fit thus unfit for medical care are neglect and death.

I am confident that there is better way to handle refractory mental impairments, honoring American individuality, integrity, and autonomy. Compassion can be combined with fiscal responsibility, allowing the mentally impaired to live. My proposed solution is the subject of my next essay. The purpose of this essay is to sound the alarm that malevolent medical reform in the Clinton Health Plan called American Health Security Act of 1993 is flourishing today in medical law. Horrors of the Progressivist eugenics of the 1920s and 1930s then paired in medicine with a perversion of Darwinism are again possible in our current American medical-legal context which honors integrity to the medical program while violating integrity of the patient, and which respects medical cost while disrespectful of physician and patient choice even if and when patient or family will pay for excellent care.

Under AHSA's legislative progeny, patients are collectivized and physicians are criminalized. Utilitarianism literally is deadly "Medically necessary" means whatever government or insurance will pay for. Psychiatric diseases only minimally responsive to treatment and chronic serious mental illness will not be treated. If limited American money, medicine, and time are invested only in inevitable medical success, then America's medicine by its medical law will be Medical Darwinism encouraging survival of the fittest by requiring extinction of the unfit.

(1) See American Health Security Act, H.R./S./ 103d Cong., 1st Sess. (1993) (hereinafter AHSA), reprinted in PRESIDENT CLINTON'S HEALTH CARE REFORM AND HEALTH SECURITY ACT AS PRESENTED TO CONGRESS ON OCTOBER 27, 1993 (Commerce Clearing House 1993); THE PRESIDENT'S HEALTH SECURITY PLAN (Random House 1993).

(2) See Madeleine E Cosman, ABCs of the Clinton Medical World, CARDODO OMNIBUS J., Apr. 1994, reprinted July, 1997, at 1, 1-8.

(3) See Toward an Urban Health Agenda, in J. URBAN HEALTH, BULL. N.Y. ACAD. MED., June 1998, at 2, 2; Bernard S. Arons et al., Mental Health and Substance Abuse Coverage Under Health Reform, HEALTH AFFAIRS, Spr. 1999, at 1, 1 (Introduction by Hillary Rodham Clinton).

(4) Health Insurance Portability & Accountability Act of 1996, H.R. 3103, 104th Cong., 2nd Sess. (1996), enacted, 42 U.S.C. (sections) 201 (1996); amended by, 42 U.S.C. (sections) 264, 1395 b-5 (1997) (hereinafter HIPAA).

(5) Balanced Budget Act of 1997, Pub. L. No. 105-33 (Aug. 5, 1997).

(6) AHSA, supra note 1, at (sections) 1115.

(7) See generally Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. (1993) (statement of Dr. Frank McArdle, Manager, Research Group, Hewitt Associates, Washington, D.C.).

(8) See text at notes 65-67, infra, and subsections entitled Serious Chronic Mental Illness, Banal Benefits, and Actuarial Equivalents.

(9) See KARL BINDING & ALFRED ROCHE, PERMITTING THE DESTRUCTION OF UNWORTHY LIFE (Felix Meiner Verlag 1920), reprinted in 8 ISSUES IN LAW & MED. 231 (Walter E. Wright, trans., and Patrick G. Derr, ed. 1992); and

NUREMBURG TRIALS; THE MEDICAL CASE. TRIAL OF WAR CRIMINALS BEFORE THE NUREMBURG MILITARY TRIBUNALS, vols. 1, 2 (U.S. Gov't Printing Office, Wash., D.C. 1948). Four important studies are: HENRY FRIEDLANDER, THE ORIGINS OF NAZI GENOCIDE: FROM EUTHANASIA TO THE FINAL SOLUTION (1994); ROBERT SAUL FRIEDLANDER, NAZI GERMANY'S CRIMES AGAINST THE JEWS (1997); ROBERT PROCTOR, RACIAL HYGIENE: MEDICINE UNDER THE NAZIS (1988); MAX WEINREICH, HITLER'S PROFESSORS: THE PART OF SCHOLARSHIP IN GERMANY'S CRIMES AGAINST THE JEWISH PEOPLE (1946).

(10) See M.P. Cosman, The Criminalization of American Medicine, 6 NAT'L TRIAL LAW. 1 (1994).

(11) See Leo Alexander, Medical Science Under Dictatorship, 241 NEW ENG. J. MED. 39 (1949).

(12) Olmstead v. United States, 277 U.S. 438 (1928) (Brandeis, J., opinion).

(13) AHSA, supra note 1, at (subsections) 1101-1115, 1122-1123, 1131-1136, 1409-1410, 1153-1154.

(14) Id. at (subsections) 3001-3601, including Subtitle E on medically under-served populations, and Subtitle F on the Public Health Initiatives Fund.

(15) Id. at (sections) 1115.

(16) Id. at Title III, especially (subsections) 3001-3601.

(17) Id. at (sections) 1115(b)(1)(A) & (B).

(18) Id.

(19) Id. at (sections) 1115(b)(1)(B)(2).

(20) Id. at (sections) 1115(e).

(21) Id. at (sections) 1115(b)(1)(B)(2).

(22) Id. at (sections) 1115(b)(3).

(23) Id. at (subsections) 1001-1004.

(24) Id. at (sections) 1115(b)(4); W.G. MANNING, ET AL., EFFECTS OF MENTAL HEALTH INSURANCE: EVIDENCE FROM THE HEALTH INSURANCE EXPERIMENT (RAND Corp. 1989); and W.G. MANNING, ET AL., USE OF OUTPATIENT MENTAL HEALTH CARE: TRIAL OF A PREPAID GROUP PRACTICE VERSUS FEE FOR SERVICES (RAND Rep. R-3277) (Nat'l Inst. of Mental Health 1986).

(25) AHSA, supra note 1, at (sections) 1115(c)(2)(D).

(26) Id. at (sections) 1115(c)(2)(A)(i)-(iii), (B), & (C).

(27) Id. at (sections) 1115(c)(2)(C).

(28) Id. at (sections) 1115(c)(2)(E).

(29) Id. at (sections) 1115(d).

(30) Id. at (sections) 1115(d)(2)(C).

(31) Id. at (sections) 1115(d)(2)(C)(i), (ii), (iii).

(32) Id. at (sections) 1115(d)(2)(C)(iii).

(33) Id. at (sections) 1115(e).

(34) Id. at (sections) 1115(e)(1).

(35) Id. at (sections) 1115(e)(2)(C)(i).

(36) Id. at (sections) 1115(e)(2)(C)(i).

(37) Id. at (sections) 1115(e)(2)(C)(ii)(E).

(38) Id. at (sections) 1115(e)(2)(E).

(39) Id. at (subsections) 1131-1136, 1371-1375, 3467, 3501, and 3701.

(40) Id. at (sections) 1115(d)(1)(D).

(41) Id. at (sections) 1115(e)(2)(D).

(42) Id. at (sections) 1115, & Part 3 (subsections) 1131-1136, & 3501.

(43) Id.

(44) Id. at (subsections) 1542 & 3461 for such "enabling services" as transport and translation.

(45) Id. at (sections) 3401, providing \$100 million dollars for each year during 1995-2000; (sections) 3501, on the Public Health Initiatives Fund; and (sections) 3467, which authorizes, and (sections) 3701, which creates the Fund.

(46) Id. at (subsections) 1153, 1329, 1542; the so-called enabling services under (sections) 3461 included transportation, community outreach,

translation services, and patient education.

(47) Id. at (subsections) 1324-1325, 1327, 1329, 1386, 1394, 1410-1411, 1413, 1504; (sections) 3511, on integration of services; and (sections) 3675, on local education agencies.

(48) Id. at (sections) 3511(b)(3).

(49) Id. at (sections) 3511(b)(8).

(50) Id.

(51) Id. at (sections) 3411, on linkages among providers, health plans, information systems, and networks; and (sections) 3511, on reports on integration of mental health services.

(52) Id. at (sections) 5102, on privacy See also HIPAA, *supra* note 4, and note 96, *infra*. My comments are in parentheses and italics. Whoever makes a false statement to a health care plan administrator is liable to a five year prison term (HR 3103 at 205-206).. (That includes patients. Psychiatrists appalled at revealing confidential information about patients pledge to destroy psychiatric records rather than subject themselves and their patients to risk of prison for the crime of maintaining confidentiality.) Failure to provide information to a criminal investigator carries a five-year prison term (HR 3103 at 206).. (What happened to physicians' protections under the Constitution's 4th and 5th Amendments?) "In ANY investigation relating to any act or activity involving a Federal health care offense.... (the Attorney General) may require the production of any records that may be relevant" (HR 3103 at 208). The person supplying patient records in response to such a subpoena "shall not be liable in any court of any State or the United States to any customer or other person for such production or for nondisclosure of that production to the customer" (HR 3103 at 210). Health information so produced cannot be used against the patient "unless the action or investigation arises out of and is directly related to the receipt of health care or payment for health care" (HR 3103 at 210). (This ends patient privacy and empowers government to use medical records to prosecute ad lib.).

(53) See INSTITUTE OF MEDICINE, BROADENING THE BASE OF TREATMENT FOR ALCOHOL PROBLEMS (Nat'l Acad. Press 1990); INSTITUTE OF MEDICINE, TREATING DRUG PROBLEMS, Vol. 1. (Nat'l Acad. Press 1990); D.P. RICE, ET AL., THE ECONOMIC COSTS OF ALCOHOL AND DRUG ABUSE AND MENTAL ILLNESS. (DHHS (ADM) 901694) (Alcohol, Drug Abuse and Mental Health Admin. 1990).

(54) An important introduction to this problem is the testimony of Dr. Crowell in 1993. Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. 3 (1993) (statement of Dr. Areta Crowell, Dir., L.A. County Mental Health Dep't).

(55) Id. at 3, 4.

(56) Id. at 4.

(57) See id. at 5.

(58) AHSA, *supra* note 1, at (sections) 1115(c)(2)(C).

(59) Id. at (sections) 1115(e)(2)(C)(i).

(60) Id. Introduction to the Health Security Act: "To ensure individual and family security through health care coverage for all Americans in a manner that contains the rate of growth in health care costs and promotes responsible health insurance practices, to promote choice in health care, and to ensure and protect the health care of all Americans, Be it enacted by the Senate and the House of Representatives of the United States of America in Congress assembled.... "See also AHSA Section 2, "Findings," listing the imperfections of the "current health care system."

(61) See W.G. MANNING, EFFECTS OF MENTAL HEALTH INSURANCE: EVIDENCE FROM THE HEALTH INSURANCE EXPERIMENT (RAND Rep. R-3815) (Nat'l Inst. of Mental Health/Health Care Finance Admin. & RAND Corp. 1989).

(62) See the reasoned testimony of Dr. Richard Frank. Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human

Resources, 103d Cong., 1st Sess. (1993) (statement of Dr. Richard Frank, Prof. of Health Econ., Johns Hopkins Univ., Baltimore, MD).

(63) Id. at 7.

(64) See generally G.N. GROB, FROM ASYLUM TO COMMUNITY (1991).

(65) See generally Crowell, *supra* note 54, and Frank, *supra* note 62.

(66) Crowell, *supra* note 54, at 5.

(67) See generally ALAN LIEBERSON, THE PHYSICIAN'S GUIDE TO ADVANCE MEDICAL DIRECTIVES (1993); P Diehr, et al., Ambulatory Mental Health Services Utilization in Three Provider Plans, 22 MED. CARE 1 (1984); A. Dill & D.A. Rochefort, Coordination, Continuity, and Centralized Control: A Policy Perspective on Service Strategies for the Chronically Mentally Ill, 45 J. SOCIAL ISSUES 145 (1989); E. Keeler, et al., The Demand for Episodes of Mental Health Care, 7 J. HEALTH ECON. 69 (1988); R.G. Frank, et al., A Model Mental Health Benefit in Private Health Insurance, HEALTH AFFAIRS, Spr. 1992, at 99, 99; R.G. Frank, et al., Paying for Mental Health and Substance Abuse Care Under Reform, HEALTH AFFAIRS, Spr. 1991, at 337, 337.

(68) State medical care experiments have generated a huge bibliography. Oregon is a good example. For a selection of views, see generally David C. Hadorn, Setting Health Care Priorities in Oregon: Cost Effectiveness Meets the Rule of Rescue, 265 JAMA 2218 (1991); David Eddy, What is Going On in Oregon? 266 JAMA 417 (1991); Michael J. Astrue, Pseudoscience and the Law: The Case of the Oregon Medicaid Rationing Experiment, 9 ISSUES IN LAW & MED. 375 (1994); Donald Cohodes, Pragmatism and Health Care Reform, HEALTH AFFAIRS, Spr. 1994, at 74.

(69) On managed competition see A.C. Enthoven, Managed Competition in Health Care Financing and Delivery: History, Theory, and Practice (address presented at the Robert Wood Johnson Foundation "Changes in Health Care Financing Initiative" Workshop (Jan. 1993). On global budgets and related cost containment methods, see generally PAUL STARR, THE LOGIC OF HEALTH-CARE REFORM (1992); THE 21 ST CENTURY AMERICAN HEALTH SYSTEM POLICY DOCUMENTS (1991); S.S. Sharfstein & A.M. Stoline, Reform Issues for Insuring Mental Health Care, HEALTH AFFAIRS, Fall 1992, at 84.

(70) Enthoven, *supra* note 68.

(71) See M. Schlesinger & D. Mechanic, Challenges for Managed Competition from Chronic Illness, HEALTH AFFAIRS, Supp. 1993, at 123, 123; and M. Schlesinger & D. Mechanic, Chronic Illness Challenges Managed Competition (unpublished manuscript presented at the Institute for Health, Health Care Policy and Aging Research, Rutgers Univ.; Dep't of Epidemiology and Public Health, Yale Univ. Med. School, Nov. 1992).

(72) Recommendations of the Little Rock Working Group on Mental and Substance Abuse Disorders in Health Care Reform 8-9 (unpublished manuscript presented at the Centers for Mental Healthcare Research, Dep't of Psychiatry, Univ. of Arkansas for Med. Sciences, Feb. 3-5, 1993) (citations omitted, emphasis added).

(73) Id. at 13 (citation omitted).

(74) In their introduction, the Little Rock Working Group expresses the altruistic "sacrifice" philosophy behind their recommendations:

Recognizing that all elements of the present health care system must share

in the sacrifice, we propose to reallocate expenditures away from the expensive, at times non-essential services, to those described in our recommendations. This requires massive change in mental and substance abuse

services where, until now, incentives have been directed toward use of inpatient services.

Id. at 7. "Reinsurance and other risk-sharing approaches (e.g., mixed capitated/fee-for-service systems) (may be necessary for) ... severe and persistent ... disorders." Id. at 13.

(75) See Frank, *supra* note 62.

(76) See generally D. Mechanic, *The Evolution of Mental Health Services and Mental Health Services Research*, in *THE FUTURE OF MENTAL HEALTH SERVICES RESEARCH* (C.A. Taube, et al., eds., U.S. Gov't Printing Office 1989); D. Mechanic, *Strategies for Integrating Public Mental Health Services*, 15 HOSP. & COMMUNITY PSYCHIATRY 797 (1991); G. Norquist & K. Wells, *Mental Health Needs of the Uninsured*, 48 ARCHIVES OF GEN. PSYCHIATRY 476 (1991).

(77) Frank, *supra* note 62, at 7-8.

(78) See AHSA, *supra* note 1, at (sections) 1115(b)(2): Case Management.

(79) Long term care is not among the mental health benefits listed in AHSA, *supra* note 1, (sections) 1115. Long term care is one of two subjects under Title II "New Benefits," Subtitle B: (sections) 2101 deals with state programs for individuals with disabilities requiring long term care; (sections) 2103 defines disabilities; (sections) 2104 lists covered services; and (sections) 2106 covers quality assurance and safeguards; (sections) 2109 provides the budget with \$4.5 billion assigned to year 1996 and \$38.3 billion to year 2003.

(80) AHSA, *supra* note 1, at (sections) 2103.

(81) Id. at (sections) 1115(d).

(82) Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. 5 (1993) (statement of Dr. Frank Docherty, Clinical Prof. of Psychiatry, Tufts Univ., Boston, MA).

(83) Id.

(84) Id. at 7.

(85) See generally R.M. Friedman & K. Kutash, *Challenges for Child and Adolescent Mental Health*, 11 HEALTH AFFAIRS, Spr. 1992, at 125, 125; Daniel B. Griffith, *The Best Interests Standard: A Comparison of the State's Parens Patriae Authority and Judicial Oversight in Best Interests Determinations for Children and Incompetent Patients*, 7 ISSUES IN LAW & MED. 283 (1991).

(86) Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. 3 (1993) (statement of Dr. Bernie Arons, Acting Director, Center for Mental Health Services, Dep't of Health and Human Serv's).

(87) Id. at 3, 4.

(88) Id. at 2-5. See also AHSA, *supra* note 1, at (sections) 3511, requiring reporting on integration of mental health systems.

(89) Bernard Arons et al., *Mental Health and Substance Abuse Coverage Under Health Reform*, HEALTH AFFAIRS, Spr. 1994, at 195, 195.

(90) Richard G. Frank, H.H. Goldman, & T.G. McGuire, *A Model Mental Health Benefit in Private Health Insurance*, HEALTH AFFAIRS, Fall 1992, at 98.

(91) Arons, *supra* note 89.

(92) Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. (1993) (statement of Senator Paul D. Wellstone, Chair).

(93) Mental Health and Substance Abuse Under the Health Security Act: Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. (1993) (statement of Honorable Mike Kopetski).

(94) Wellstone, *supra* note 92, at 4.

(95) Mental Health and Substance Abuse Under the Health Security Act:

Hearings on the American Health Security Act Before the U.S. Senate Comm. on Labor and Human Resources, 103d Cong., 1st Sess. 6-8 (1993) (statement of Dr. Frank McArdle, Manager, Research Group, Hewitt Associates, Wash., DC).

(96) See HIPAA *supra* notes 4 & 52. Draconian provisions of AHSA now are the law of the land via HIPAA. In May, 1996, HR 3103 and S 1028 moved successfully through Congress, passing the Senate 100 to zero. Taken almost verbatim from AHSA, about 100 pages of criminal sanctions and penalties against physicians, surgeons, and patients make a practitioner strictly liable, guilty even with no intent to do the forbidden deed. Forbidden "crimes," e.g., errors in coding for reimbursement, providing "medically unnecessary" care, and "referrals" prosecuted under the Stark Laws, are punishable by the formidable "Three Fs": guilt of a felony, fines up to \$25,000 per incident, and five years in federal prison.

Here are seven examples (with my comments in parentheses and *italics*): (1) The Health Care Fraud and Abuse Trust Fund collects criminal fines, civil monetary penalties, and forfeited property from physicians and surgeons at conviction or before conviction. "The court, in imposing sentence on a person convicted of a Federal health care offense, shall order the person to forfeit property, real or personal, that constitutes or is derived, directly or indirectly, from gross proceeds traceable to the commission of the offense" (HR 3103 at 211). (2) Rewards are offered to patients and to medical staff members to act as informants or whistle-blowers who share in money penalties imposed on physicians (HR 3103 at 161). (These are qui tac actions under the False Claims Act of 1986). (3) Civil monetary penalties are \$10,000 for each instance of "failure to comply with statutory obligations" which include incorrect coding and medically unnecessary service (HR 3101 at 195). (The physician is liable (a) for every billing secretary's keystroke and (b) for exercising medical judgment that dares to differ from the third party payer insurance company which, if not wanting to pay, will deem the procedure medically unnecessary.) (4) Forbidden "remuneration" to doctors includes "transfers of items or services for free or for other than fair market value" (HR 3101 at 198). (Charity and friendly favors are considered criminal fraud). (5) No proof of specific intent to defraud is necessary (HR 3101 at 200). (This is strict liability for crime.) (6) A "Federal health care offense" is defined as any violation involving "any public or private plan or contract, affecting commerce, under which any medical benefit, item, or service is provided to any individual" (HR 3101 at 202). (The language of HR 3101 at this point is so over-inclusive that any innocent doctor can stimulate the ire of an overzealous investigator). (7) "Defrauding" any health care benefit program carries a fine and/or imprisonment for ten years, or life imprisonment "if the violation results in death" (HR 3101 at 203). (A physician treating a terminally ill patient with "excessive care" could be jailed for life. Several physicians are serving in federal penitentiaries for medically unnecessary medicine and surgery. See, e.g., United States v. Rutgard, No. 96-50309 (9th Cir. filed Mar. 6, 1997) (reversing a criminal forfeiture order in the amount of \$7,564,441.22 against Dr. Rutgard), and United States v. Anderson, 85 E Supp. 2d 1084 (D.Kan. 1999) (following a nine-week trial, a jury convicted two doctors and the head of a medical facility of Medicare kickback offenses in connection with an alleged bribery scheme by the doctors, who referred patients to the facility in exchange for compensation)).

(97) McArdle, *supra* note 7.

(98) *Id.* at 11.

(99) AHSA, *supra* note 1, at Title III, Subtitle A defines work-force priorities under federal payments. Subtitle B demonstrates the micro-managing of academic health centers, already underway because of HIPAA and other laws, requiring that after five years, no matter the physicians' interests and talents, 50% of all new physicians must be

trained in primary care rather than specialties. This increases primary care residency positions by 7% each year for five years and decreases specialty training positions by 10% annually. The only medical specialties allowed to grow are family medicine, general internal medicine, and general pediatrics. Any medical school and university center not obeying those guidelines gets no government money, and the Council on Graduate Medical Education determines which residencies are to be accredited, (thus allowed to exist, and which not). The Council reserves internships, residencies, and fellowships for members of under-represented minority groups. Under Subtitle C's health research initiatives, government will fund scientific studies of cost effectiveness of care, administrative simplification (as in national data banks of medical records), practice guidelines, and consumer choice. Subtitle D discusses core functions of public health programs and preventive medicine called "preventive health," while Subtitle E is devoted to the medically underserved populations. Subtitle F is on mental health and substance abuse.

(100) McArdle, *supra* note 7, at 21.

(101) Id. at 17.

(102) See generally Donald Cohodes, *Pragmatism and Health Care Reform*, HEALTH AFFAIRS, Spr. 1994, at 264.

(103) Id. at 18.

(104) Id.

(105) Id.

(106) Id. at 8 (emphasis added).

(107) Id.

(108) See generally Stanley Herr et al., *No Place to Go: Refusal of Life-Sustaining Treatment by Competent Persons with Physical Disabilities*, 8 ISSUES IN LAW & MED. 3 (1992).

(109) See generally *Is THERE A DUTY TO DIE?* (J.M. Humber et al. eds. 2000); Jerome C. Arnett, *Is There a Duty to Die?*, 5 MED. SENTINEL 183 (2000); J. Hardwig, *Is There a Duty to Die?* 27 HASTINGS CENTER REP. 34 (1997); Richard Lamm, Governor of Colorado, *Address to Colorado Health Lawyers' Association*, N.Y. TIMES, Mar. 29, 1984, at Al.

(110) See generally David C. Hadorn, *Setting Health Care Priorities in Oregon: Cost Effectiveness Meets the Rule of Rescue*, 265 JAMA 2218 (1991); David Eddy, *What is Going On in Oregon? 266 JAMA 417 (1991); Michael J. Astrue, *Pseudoscience and the Law: The Case of the Oregon Medicaid Rationing Experiment*, 9 ISSUES IN Law & MED. 375 (1994); and Donald Cohodes, *Pragmatism and Health Care Reform*, HEALTH AFFAIRS, Spr. 1994, at 74.*

(111) See notes 9 and 11 *supra* and accompanying text.

(112) 274 u.s. 200 (1927).

(113) Id. at 207 (emphasis added). The following puts the quotation in context:

We have seen more than once that the public welfare may call upon the best

citizens for their lives. It would be strange if it could not call upon those who already sapped the strength of the state for these lesser sacrifices, often not felt to be such by those concerned, in order to prevent our being swamped with incompetence. It is better for all the world

if instead of waiting to execute degenerate offspring for crime, or to let them starve for their imbecility, society can prevent those who are

manifestly unfit from continuing their kind. The principle that sustains compulsory vaccination is broad enough to cover cutting the fallopian tubes. Three generations of imbeciles are enough.

Id. (citation omitted, emphasis added).

(114) 316 U.S. 535 (1942).

(115) See generally DAVID ROTHMAN & SHEILA A. ROTHMAN, THE WILLOWBROOK WARS (1984).

(116) See generally JAY KATZ, EXPERIMENTATION WITH HUMAN BEINGS (1972).

(117) Kaimowitz v. Michigan Dep't of Mental Health (Cir. Ct., Wayne Co., Mich. 1973).

(118) See generally ALAN MEISEL, THE RIGHT TO DIE, 2 VOLS. (1995); and compare the entire back run of this journal, ISSUES in Law & Medicine, especially Daniel Avila, Medical Treatment Rights of Older Persons and Persons with Disabilities: 1991-92 Developments, 8 ISSUES IN LAW & MED. 429 (1993); his Medical Treatment Rights of Older Persons and Persons with Disabilities: 1992-93 Developments and Emerging Trends, 9 ISSUES IN LAW & MED. 345 (1994); Diane Coleman, Withdrawing Life-Sustaining Treatment from People With Severe Disabilities Who Request It: Equal Protection Considerations, 8 ISSUES IN LAW & MED. 55 (1992); and Mark E. Haddad, Cruzan and the Demands of Due Process, 8 ISSUES IN LAW & MED. 205 (1992).

(119) 760 S.W.2d 408 (Mo. 1989) (en banc); affirmed sub. nom. Cruzan v. Director, Missouri Dep't of Health, 497 U.S. 261 (1990).

(120) In re Quinlan, 70 N.J. 10, 355 A.2d 647 (1976), cert. den. sub. nom. Garger v. New Jersey, 429 U.S. 922 (1976).

(121) Washington v. Harper, 494 U.S. 382 (1990); Mills v Rogers, 457 U.S. 291 (1982); Bouvia v. Superior Court, 179 Cal. App. 3d 1127, 225 Cal. Rptr. 297 (1988); Bartling v. Superior Court, 163 Cal. App. 3d 186, 209 Cal. Rptr. 220 (1984); Matter of Storar, 438 N.Y.S.2d 266 (Ct. App. 1981); Schloendorff v. Society of New York Hospital, 211 N.Y. 125, 105 N.E. 92 (1914); Blackburn v. State, 23 Ohio St. 146 (1873).

(122) See generally Nancy J. Osgood & Susan A. Eisenhandler, Gender and Assisted and Acquiescent Suicide: A Suicidologist's Perspective, 9 ISSUES IN LAW & MED. 361 (1994).

(123) See generally Paul Steven Miller, The Impact of Assisted Suicide on Persons With Disabilities -- Is It A Right Without Freedom? 9 ISSUES IN LAW & MED. 47 (1993); Peter A. Ubel, Assisted Suicide and the Case of Dr. Quill and Diane, 8 ISSUES IN LAW & MED. 487 (1993).

(124) See, e.g., Nancy J. Osgood, Assisted Suicide and Older People -- A Deadly Combination, 10 ISSUES IN LAW & MED. 415 (1995):

On June 4, 1990, fifty-four-year-old Janet Adkins ended her life lying on a cot in the back of a Volkswagen van parked in a Michigan suburb. Aided by a retired pathologist, Dr. Jack Kevorkian, Adkins was hooked up to his homemade "suicide machine." She had a needle inserted in her arm, which first started saline flowing and, then, when she pressed the button on the macabre death machine, sent first a sedative and then deadly potassium chloride flowing into her veins. An active woman with loving children and

grandchildren, Adkins has flown two thousand miles from her Oregon home to

Michigan to seek Kevorkian's assistance in ending her life when she was diagnosed with Alzheimer's disease.... She made a deliberate decision to end her life rather than face the mental decline associated with senile dementia.

Id. at 415 (emphasis added).

(125) See generally David Hadorn, *Setting Health Care Priorities in Oregon: Cost Effectiveness Meets the Rule of Rescue*, 265 JAMA 2218 (1991). Madeleine Pelman Cosman, Ph.D., Esq., Attorney and president of Medical Equity, Inc., San Diego, Cal., a national medical and law practice consultancy; B.A., Barnard College, 1959; M.A., Hunter College, 1960; Ph.D., Columbia University, 1964; J.D., Cardozo School of Law, 1995. Dr. Cosman is Professor Emerita of the City College of City University of New York where for twenty-eight years she taught medical students medical law, medical business, and medical history. She is a life Fellow of the New York Academy of Medicine. She is a member of the New York Bar, New Jersey Bar, and a Barrister of the American Inns of Court. One of her fifteen books was nominated for the Pulitzer Prize and National Book Award.

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Vatical settlements: a new way to nursing home private pay.

Zadoff, Michael

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Cash-strapped residents still have an option before turning to Medicaid

Vatical settlements, a financial resource for the terminally or chronically ill, have been around for more than a decade. But most people know little, if anything, about them. For residents of senior congregate living facilities, which include nursing homes, assisted living facilities and independent living communities, they can be a lifeline - the answer to their needs for immediate cash for a variety of reasons. These can include upgrading their living quarters to a private room, paying for costly experimental treatments not covered by traditional insurance plans or giving financial relief to family members.

Vatical settlements allow individuals with terminal or chronic illnesses to sell their life insurance policies for cash. They first became

a financial planning tool in the mid-1980s in response to the AIDS epidemic. Viaticals changed the way we view insurance policies. Once regarded only as a benefit to surviving family members, life insurance policies soon became viewed as an existing asset, much like stocks and bonds, real estate and savings accounts.

Viaticals were particularly appropriate for People With AIDS (PWAs) because they had a life-limiting illness, which, at the time, had a very predictable term. In many cases, PWAs had limited access to medication and hospital care because they lost their jobs and health benefits when they became ill. Many died destitute. In recent years, viaticals have become more mainstream and are benefiting people with a broader range of life-limiting illnesses. In addition to PWAs, they include people of all ages with terminal illnesses, as well as residents of senior congregate living communities. Viatical settlements are becoming increasingly popular among seniors who require immediate cash for a wide range of needs.

Since most residents and their families are probably unfamiliar with viatical settlements, it is in their best interests - and those of management, social workers and others in direct contact with residents - to make them aware of this financial resource. Financial planning counsel can be just as important to a resident's well-being as the attention given to his medical, social, psychological and intellectual needs.

A viatical settlement is a way to access cash and reduce financial strains and resulting emotional unrest for both the resident and family. It makes the life insurance policy a valuable financial asset that can improve the quality of the resident's remaining years.

The following is a basic overview of viatical settlements and the key questions that management and personnel dealing directly with residents might want to address.

What is a viatical settlement?

The word "viatical" comes from the Latin word "viaticum," which refers to the necessary money or supplies given to a person embarking on a long or difficult journey. In the Christian church, "viaticum" is the blessing of the Eucharist given to provide spiritual sustenance to a dying person or one in danger of death.

A viatical settlement is a contract that allows an individual with a relatively short life expectancy to sell a life insurance policy for cash to a third party - an individual or business entity - at a discounted value of the policy's face value. Simply, the person gets cash in exchange for relinquishing the death benefits of the life insurance policy to the person paying for it, less a certain percentage of the benefit. The amount that the viator - the seller of the policy - receives depends upon his/her life expectancy. The viator can get as much as 80% of the policy's face value. Basically, the less time they have left to live, the more money they get.

By viewing a life insurance policy as a current asset, the insured can now access funds - the death benefits - virtually immediately as a living benefit.

The elderly might need the cash for a variety of reasons, although the needs of those with life-limiting conditions are often quite basic, but costly. Having these needs met can reduce the older person's emotional strains and subsequently improve his or her health. These needs could include:

- * Paying for experimental treatments not covered by traditional medical insurance policies;
- * Paying for nursing home or assisted living community care if family members are footing the bill;
- * Moving to a private room, perhaps a larger one with more luxuries and conveniences, such as a private phone;
- * Having a private nurse;
- * Paying travel expenses for family visits to the facility;
- * Funding educational costs for a grandchild;

* Making a gift to a loved one.
Nearly every type of life insurance policy qualifies for a viatical settlement, providing it's been in force for at least two years. The process is simple:

To determine the life expectancy of the viator, the viatical settlement company has a board of physicians evaluate the person's illness, basing their findings on medical records, laboratory reports and current actuarial tables. The viator then receives the cash payment after transferring ownership of the life insurance policy to the viatical settlement provider. The entire process takes about a month. The policy is then maintained by the viatical settlement company, which also pays the premiums.

What about taxes?

The federal government has established strict guidelines governing the viatical industry. Most significant is the 1996 Health Insurance Portability & Accountability Act which exempts viators from paying federal income tax on these settlements if they are terminally or chronically ill. The Act defines a terminally ill person as one with a medically certified life expectancy of less than 24 months. A chronically ill person is defined as one who is unable to perform at least two activities associated with daily living. Only in these qualified situations are viatical settlements tax-free. Furthermore, individuals qualifying for this tax-free benefit must use a viatical settlement provider who is licensed in the state where they live. In some states, funds are also exempt from state taxes.

Are viaticals for the healthy?

Now, anyone over the age of 70 with a life insurance policy who is in serious need of money can qualify for what is called a "senior settlement," an increasingly popular option. These people may be perfectly healthy or may have suffered a heart attack or stroke or have other ailments they can live with indefinitely. However, given their age and statistical data, they are candidates for such a settlement, based on actuarial life expectancy tables established by insurance companies. The shortcoming here is that a senior settlement is not tax-free.

What are the risks?

The risks for the family are clear: The children or other survivors are no longer the beneficiaries of the insurance policy. They've waived their rights to the proceeds. It has been sold and the buyer, who's now paying the premiums, will receive the death benefits. It's up to the resident and family to decide. A viatical settlement is but one financial option, but it is one that can provide convenient access to much-needed cash.

Michael Zadoff is president and founder of Dedicated Resources, Inc., a viatical settlement company in Delray Beach, Florida. Founded in 1988, it is one of the oldest viatical settlement companies in the United States. For further information, phone: (800) 677-5026; fax: (561) 495-9089; or send e-mail to: drviatical@aol.com.

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Self-Funded Plans Face More Legislative Threats.

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ED UEECK IS A MANAGING DIRECTOR WITH PACIFIC RISK MANAGEMENT SERVICES, FOUNTAIN VALLEY, CALIF. In 1994, sponsors of self-funded employee health plans and their advisers watched Congress debate a single piece of federal legislation that could have altered the entire U.S. health care system. Today, pending state and federal bills appear to have the potential to drastically alter health care one bill at a time. Sponsors of self-funded plans must also look out for the effects of existing laws that give the Internal Revenue Service, the U.S. Department of Health and Human Services and the U.S. Department of Labor the power to write and enforce health plan regulations. Major federal laws that affect self-funded plans include the Health Insurance Portability, Accountability Act, also known as Kassebaum-Kennedy Act, the Mental Health Parity Act, and the Employee Retirement Income Security Act, ERISA, the legislation that most significantly affects self-funded plans, was signed into law in 1974. ERISA has helped many employers provide affordable health coverage by exempting self-funded plans from offering the state mandated benefits that fully insured plans must offer. In addition, self-funded plans' exposure to damages from lawsuits is currently limited to actual, not punitive, damages. Actions brought against self-funded plans are resolved in federal courts, which are somewhat more insulated from political interests than the state courts. Many of these protections may be eroded by the consumer backlash against health maintenance organizations. For example, Texas recently passed legislation that allows participants to sue managed care plans for malpractice. While the bill was primarily aimed at HMOs, it could have significant ramifications for self-funded plans if the state court decides that plan service providers or employers themselves are doing more than providing indemnification through a plan of benefits. Texas courts must quickly clarify the difference between providing benefits under a plan and practicing medicine. Other states have, or are considering, legislation that would set the minimum level of medical excess loss coverage. For example, some states want to quantify what is and isn't a self-funded plan by establishing rules for the stop-loss level a plan can have. Self-funded plans buy stop-loss coverage to protect themselves from unexpected increases in use or catastrophic claims. There are two primary types of stop-loss coverage: specific, which protects the plan against a single catastrophic loss, and aggregate, which generally limits a plan's exposure to large fluctuations caused by poor plan design, economic changes or random variations that lead to unexpected increases in large claims below the specific deductible. Under the ERISA interpretations proposed in some states, if a plan had specific stop-loss levels of less than \$10,000 or aggregate coverage of less than 125 percent, it would not be considered self-funded and would be subject to mandated benefits as outlined in the state law. Maryland's attempt at quantifying what constitutes a self-funded plan failed in federal district court, and on appeal was not reviewed by the U.S. Supreme Court. However, as states continue to regulate the small group market, there will probably be more attempts in this direction. HIPAA is supposed to ensure that people can get insurance quickly if they change jobs, even if they have a preexisting medical condition. It also creates a

pilot program for medical savings accounts and provides tax benefits for self-employed individuals who purchase health insurance. But the bill contains a myriad of additional features that directly affect self-funded plans. For example, employees must not allow a gap in coverage of longer than 62 days in order to be entitled to waive preexisting condition exclusions when joining their new employers' health plans. Employers must provide certificates of prior coverage so that departing employees can show new employers they are eligible for coverage without any preexisting condition exclusions. However, in the few short years since HIPAA was passed, employers have seen a marked increase in the amount of fraudulent certificates. This has created a need for anti-fraud measures and further increases costs to the health plan. The Mental Health Parity Act Under the Mental Health Parity Act of 1996, employers that offer either fully or self-funded health plans with mental health benefits may not set different lifetime dollar limits or annual dollar limits for the mental health benefits. Many benefits consultants, consumer advocates and industry analysts view MHPA as a "skeleton" of a bill. There are many provisions in the bill that have not been clearly stipulated. The concern by many is that if states have problems implementing the MHPA, that will put pressure on the federal government to "add meat" to the skeleton. Patient Rights Patient rights appears to be one of the most volatile health care issues before Congress today. Both the Republicans and the Democrats have proposed their own approaches to protecting patients' rights. Provisions in some of the proposed bills could increase the cost of health care benefits for self-funded plans by opening the door for an increased number of lawsuits.

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